STUDIES OF CEREBRAL PALSY IN THE CHILDHOOD POPULATION OF EDINBURGH.

by

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Dedicated

To my wife and parents.

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The initial studies of cerebral palsy in Edinburgh were made during the tenure of a George Guthrie Research Fellowship in the Department of Child Life and Health in the University of Edinburgh in 1952 and 1953. Professor Ellis suggested the investigation and gave me every encouragement and stimulation during it. I owe him most grateful thanks.

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Index.

	Page.
Acknowledgements. Dedication.	
Introduction.	I.
Section I. The Definition and Classification of Cerebral Palsy in Childhood. Chapter I.	4.
Section 2. The aims, scope and methods of a survey of Edinburgh children suffering from cerebral palsy carried out in I952-53. Chapter I.	49.
Section 3. A review of the literature on the different forms of cerebral palsy and a description of the findings in 208 patients.	
Chapter I. Review of the literature on the aetiology of congenital hemiplegia.	62.
Chapter 2. Review of the literature on the aetiology of acquired hemiplegia.	75.
Chapter 3. Review of the literature on the clinical findings in hemiplegia in childhood.	II6.
Chapter 4a. Aetiological studies of 75 children suffering from hemiplegia. Introduction and Family History, The maternal factor.	I35.
Chapter 4b. The birth histories of patients suffering from congenital hemiplegia.	I72.
Chapter 4c. The aetiological findings in patients with acquired hemiplegia.	I97
Chapter 4d. The aetiological studies of hemiplegia of unknown origin.	242.
Chapter 4e. Clinical findings in hemiplegia.	248.
Chapter 5a. The literature on bilateral hemiplegia.	3I4.
Chapter 5b. Aetiological studies of bilateral hemiplegia.	3I9.
Chapter 5c. The clinical findings in patients with bilateral hemiplegia.	33I.
Chapter 6. The literature on diplegic paralysis.	342.
Chapter 7a. Family History in 79 patients suffering from diplegia. The maternal factor.	369.
Chapter 7b. The birth histories of diplegic patients.	4I4.
Chapter 7c. Consideration of the significance of abnormalities of pregnancy, labour and delivery in patients with congenital diplegia.	449.
Chapter 7d. The clinical findings in 79 cases of diplegia.	483.
Chapter 8a. Review of the literature on ataxia and ataxic diplegia.	545.

Chapter 8b. Family history of patients suffering from ataxic diplegia. The maternal factor. 561.
Chapter 8c. The birth histories of patients with ataxic diplegia. 571.
Chapter 8d. Clinical features of ataxic diplegia. 585.
Chapter 9a. Family History of patients with ataxia. The maternal factor. 604
Chapter 9b. Birth histories of patients with ataxia. 612.
Chapter 9c. The clinical findings in 15 cases of ataxia. 622.
Chapter 10. The literature on dyskinesia in childhood. 645.
Chapter 11a. Family history of patients with dyskinesia. The maternal factor. 675.
Chapter 11b. Birth histories of children with dyskinesia. 682.
Chapter 11c. The clinical findings in 17 patients suffering from dyskinesia. Unclassified cerebral palsy. 700.
Section 4. The prevalence of cerebral palsy and its social aspects in Edinburgh. 750.
Chapter 1. The prevalence of cerebral palsy amongst children in the City of Edinburgh and the problem of cerebral palsy in the community. 750.
Chapter 2. Social conditions of patients suffering from c cerebral palsy. 786.
Section 5. The association of birth injury with cerebral palsy.
Chapter 1. The association of birth injury with cerebral palsy. 827.
Chapter 2a. Birth injury as a cause of cerebral palsy. 853.
Chapter 2b. Birth injury. The criteria and causes of hypoxic birth injury. 857.
Chapter 2c. Trauma in labour and delivery. 893.
Chapter 2d. Toxic agents in parturition. 921.
Chapter 3. The dangers of parturiton in the patients suff ering from cerebral palsy in the present series. 938.
Section 6. Congenital malformations as a cause of cerebral palsy.
Chapter 1. Congenital malformations as a cause of cerebral palsy. 959.
Chapter 2a. A study of the importance of developmental malformation amongst patients su ffering from congenital cerebral palsy in the present series. 975.
Chapter2b. Social factors in developmental malformations and in the patients in the present series. 980.

Chapter 2c. The family history in cases of congenital malformation and in patients in the present series. 991.

Chapter 2d. The maternal factor in congenital malformations and in cerebral palsy. 1013.

Chapter 2e. The child with congenital malformations. 1030.

Chapter 3. Conclusions. 1043.

References. Bibliography. 1052.

Introduction.

This thesis is the result of an investigation of the prevalance, clinical findings and aetiology of cerebral palsy in the childhood population of Edinburgh which was carried out during 1952 and 1953, whilst the author held a George Guthrie Research Fellowship from the University of Edinburgh.

The aims of the investigation were, firstly to establish the prevalence of cerebral palsy in the childhood population of the city; secondly to study the clinical features of cerebral palsy and their effects on the patient's way of life; thirdly to define some of the important aetiological factors in cerebral palsy in a representative group of children in the community.

During the investigation it became increasingly apparent that the currently defined categories included in "Cerebral Palsy" did not allow for an accurate classification of cases by neurological findings. Eventually a new classification on the basis of neurological syndromes was evolved. This classification will be described and compared to previous classifications in Section 3.

It was possible to establish figures for the prevalence of cerebral palsy in the childhood population of Edinburgh, though a complete ascertainment of all patients was not made. The clinical features of cerebral palsy in the childhood community were studied and are described in Section 4. During the /

the survey it became increasingly apparent that "Cerebral Palsy" was no clinical entity. Rather it comprised a number of neurological disorders in which the only common factor appeared to be that there was motor dysfunction due to abnormality of the brain which was present in early life. The clinical features varied widely from category to category. The ways in which patients were handicapped and the extent to which they were prevented from taking part in everyday activities were very different. A detailed study was made of the clinical findings and handicaps of patients and they were compared to those described by previous authors. Thus, some idea of the importance of cerebral palsy in the community was obtained, (Section 5).

Aetiological factors which were important in one form of cerebral palsy were found to be much less important in others. Many different "causes" of cerebral palsy were found which varied from developmental malformation to traumatic head injury, and from abnormal parturition to the complications of infectious diseases in early life. The multiplicity of aetiological factors in single categories and even single patients was impressive. For example, within the category of "Ataxic Diplegia" patients were found whose disorder appeared to be genetically determined, and patients who were suffering from the effects of birth injury, parainfectious encephalomyelitis or meningitis. To take account of the multiplicity of aetiological factors it was necessary to study the heredity and social backgrounds of patients as well as their individual birth /

birth and later histories.

The current concept of cerebral palsy as being due predominantly to the effects of birth injury is a misleading simplification of the true position. In the same way as there are many different causes of stillbirth and infant death, so there are many causes of cerebral palsy in children who survive. The later sections of this thesis are concerned with demonstrating that the aetiological factors in cerebral palsy are as complex as those involved in infant mortality. Social, genetic, obstetric and many unknown factors play a part. An attempt has been made to define the importance of some of them in Sections 5 and 6.

At the expense of considerable repetition an attempt has been made to make each section of this work complete within itself. The evidence bearing on each aspect studied has been summarised in tables placed in the relevant sections whenever possible.

SECTION I.

The Definition and Classification of Cerebral Palsy in Childhood.

The Definition and Classification of Cerebral Palsy in Childhood.

Definition.

For the purposes of this thesis, cerebral palsy is used as an inclusive term to describe a group of motor disorders of young children in whom full use of the limbs is prevented by paresis, involuntary movement or inco-ordination which result from damage to the brain. Motor disturbances which are the result of progressive disease, or which are transient are excluded. Motor disorders which are primarily the result of disease of the spinal cord are also excluded.

This definition is compatible with the majority of definitions made by recent writers on cerebral palsy. (Wyllie, 1951; Benda, 1952; Andersen, 1954; Courville, 1954; Perlstein, 1955; Crothers, 1951). Thus, Polani (1956) gives as his definition, "Infantile cerebral palsy can be defined as a syndrome of non-progressive brain damage, caused by factors operating on an immature nervous system, manifestating itself at birth or in early postnatal life, showing essentially a non-fitful disturbance of voluntary movements, and frequently revealing associated handicaps - intellectual, convulsive, sensory, emotional and specific educational". Kurland (1957) states "The term cerebral palsy refers to those disorders of unknown aetiology arising during the perinatal period, that is, those affecting the motor function of the brain in which the dysfunction is recognised prior to the end of infancy (two years) ".

Disease entities such as congenital syphilis with resulting paralysis, those of a progressive nature such as tuberous sclerosis, Tay Sachs, cerebral macular degeneration, or well recognised symptom compleses which may affect motor function such as hydrocephalus, mongolism or encephalodysplasia, are not included in this cerebral palsy category."

It will be seen at once that the inclusion of "well-recognised symptom compleses which may affect motor function", in the last definition allows considerable variation in the range of disorders which are included by different workers. What are considered to be "progressive disorders" will also vary with individual clinicians. In a proportion of patients it may be very difficult to decide how much of the motor disability is the result of brain and how much of spinal cord disease.

Since the definition of "Cerebral Palsy" is essentially a clinical one, other variations in its usage are inevitable. The common factor in all disorders included in the definition is that there is paresis, inco-ordination or involuntary movement the result of brain damage, (not necessarily cerebral). The majority of patients have other clinical manifestations. The commonest of these are mental impairment and epilepsy. When the interference with motor function is mild and mental defect or epilepsy severe, it is likely that some workers will classify patients as suffering /

suffering from cerebral palsy, and others will call them epileptic or mentally defective.

It is important that the arbitrary nature of the clinical disorders which are termed "cerebral palsy" should be realised. As emphasised by Freud (1897), "cerebral palsy" is not a clinical entity. It is a collection of non-progressive disorders of the nervous system found in early childhood, which share abnormalities of voluntary movement due to brain damage as their common clinical finding. As Freud hoped would happen, some of the clinical syndromes found within the category of cerebral palsy have become well defined in aetiological, pathological and clinical terms. There remain many less frequently occurring forms of cerebral palsy about which much less is known.

The Classification of Cerebral Palsy.

Historical. The currently used classifications of cerebral palsy are based on clinical findings. They have evolved gradually and have become established only after other methods of classification have been found wanting. A brief review of the way in which the knowledge of cerebral palsy has increased and methods of classification have changed explains many of the differences and difficulties in terminology and classification which prevail today.

Initially, nervous disease in childhood was regarded as a phenomenon of nature, rather than as a subject meriting close study or scientific investigation. The prevailing attitude /

attitude to these abnormalities until the eighteenth century seems well expressed in the remark of Heraclitus of Ephesus, "Zeus amuses himself". In fact the earliest extant descriptions of children with paralysis are by artists, not authors. The hemiplegic boy in "The Transfiguration" by Raphael and "The Beggar Boy", by Ribera show extremely accurate observation of postures adopted by children with cerebral palsy.

The first account of the scientific investigation of patients suffering from cerebral palsy is probably the description of three cases of cerebral hemiatrophy by Morgagni (1762). He observed in passing that the children had suffered paralysis of one side of the body during life. He attributed the abnormality of the brain to failure of development, similar except in degree to that which was responsible for anencephaly. His clinical and pathological description of "The idiot boy of Bologna" must be another of the earliest attempts to relate pathological and clinical findings.

The age of classification by pathological findings. The chief interest of the early investigators was in pathology and clinical descriptions of cerebral palsy lagged behind the pathological. From the beginning of the nineteenth century, cerebral atrophy was extensively studied. Localised atrophy and hemiatrophy were observed to be associated with hemiplegia in childhood as early as 1827 by Cazauvielh. He presented the first full clinical description of hemiplegia in childhood and /

and recognised that epilepsy and mental defect were frequently found. His major interest, however, was to attempt to distinguish between atrophy, the result of failure of development, "de vices primitives", from atrophy due to disease acquired before, during, or after birth.

During the next thirty years there were progressively more detailed descriptions of the various forms of cerebral atrophy, (Duges, 1826; von Lallemand, 1834; Henoch, 1842; Cotard, 1868; Cruvehlier, 1862). As a result of their work it became possible to distinguish in many cases between atrophy complicating old haemorrhagic lesions, atrophy accompanied by loss of cerebral substance which usually followed cerebral softening, and atrophy which resulted from inflammatory disease. It also became apparent that the association of cerebral atrophy was by no means constant. A proportion of patients with hemiplegia were later found to have other cerebral abnormalities, and many patients with cerebral atrophy showed other, or no clinical abnormalities.

Though a number of later authors attempted to construct clinico pathological syndromes of lobar sclerosis and porencephaly, these were never convincingly successful, (Richardiere, 1885; Brissaud, 1896). They were subjected to devastating criticism by Freud (1897). He pointed out that it was unsound for a clinician to classify on the basis of pathology which his findings led him to assume was present. On the other hand, pathologists found great difficulty in deducing clinical manifestations or aetiological agents from their /

their neuropathological findings. In many cases they were dealing with the end results of healing processes in the brain which gave little indication either of the original cause or extent of brain damage. Following Freud's work, few attempts have been made to classify cerebral palsy on the basis of neuropathological findings.

Early attempts to classify by clinical findings.

Though pioneer efforts had been made by Delpech to describe the differences between what would now be called upper and lower motor neurone lesions, it was not until von Heine presented a differential diagnosis of spinal paralysis, that the importance of this was generally accepted. Delpech had described symmetrical paralysis, predominantly of the lower limbs in childhood which did not appear to be due to Pott's disease. He thus presented the first full clinical description of diplegia. von Heine suggested that the paralysis in this condition was probably cerebral in origin, and cited the frequent occurrence of mental defect and epilepsy amongst patients in favour of this. (Delpech, 1830; von Heine, 1860).

The classical papers of Little appeared in 1843 and 1862. They were little known by continental workers and their importance in shaping contemporary ideas about cerebral palsy have probably been overestimated by later English speaking authors. Little's contribution was almost entirely clinical. He was the first to demonstrate the close /

close relationship between abnormalities of parturition and paralysis in childhood. He described three types of paralysis, "Hemiplegic Rigidity", "Paraplegic or Generalised Rigidity" and a form characterised by "Disordered movements". "Hemiplegic Rigidity" was only later realised to be the same as congenital hemiplegia. (Charcot bewailed the fact that he never encountered a case of hemiplegic rigidity). Generalised and paraplegic rigidity became known as "Little's Disease" and later, "Diplegia", (Freud, 1893). His category of patients with disordered movement was ignored at the time. Little laid little emphasis on distinguishing between these clinical syndromes. He was more concerned to stress their common origin in abnormalities of parturition.

After the publication of Little's papers, the clinical study of patients suffering from cerebral palsy was accelerated, and more attention was paid to classification. In France, Charcot and his disciples were especially interested in the correlation of clinical and neuropathological findings. (Charcot, 1886; Brissaud, 1894a, 1894b; Cotard, 1868; Marie, 1888; Jendrassik and Marie, 1885; Richardiere, 1885). There was much controversy in France and Germany as to whether or not paraplegic and generalised rigidity could be regarded as manifestations of the same disease entity differing only in extent and severity. Some authors, notably Charcot himself, and Dejerine, maintained that paraplegic rigidity was due to disease of the spinal cord, whereas generalised rigidity was due to cerebral abnormalities.

Degerine /

(Dejerine, 1905). The majority of authors considered them to be similar in nature and underlying pathology, being due to abnormalities of the brain. (Freud, 1893; Ganghofner, 1894, 1895).

In England the same controversy existed but was much less intense. The emphasis was on the accurate description of the neurological abnormalities shown by patients suffering from the different types of cerebral palsy and on their associated abnormalities. The classification in general use was essentially that of Little. The categories which were usually recognised were hemiplegia, paraplegic rigidity, generalised rigidity and patients showing involuntary movements as their predominant motor disability. (Ross, 1882; Hadden, 1884; Gowers, 1888).

In the United States the term generalised rigidity was not much used. In 1885 Sarah McNutt attributed most cases of congenital cerebral palsy to subdural haemorrhages, the result of birth injury. When the haemorrhage was unilateral it caused hemiplegia. When it was bilaterial it caused "Bilateral hemiplegia". This term was used by most American writers and included cases which would have been diagnosed as suffering from "Generalised rigidity" in Europe. (Osler, 1889; Lovett, 1888; Sachs and Peterson, 1890). Thus, American classification consisted of -

Hemiplegia
Spastic paraplegia
Bilateral Hemiplegia
Cases showing involuntary movements as their main disability

The /

The latter were not always clearly distinguished from those with "Bilateral Hemiplegia".

The more detailed study of patients showing involuntary movements from shortly after birth as their chief motor disability resulted in the definition of another category of cerebral palsy. (Clay Shaw, 1873; Weir Mitchell, 1874; Raymond, 1876; Ollmont, 1878; Audry, 1892; Gowers, 1876). The descriptive term given to it was "Athetosis" which had been coined by Hammond in 1871 to describe the involuntary writhing movements of the fingers observed in some patients with hemiplegia. In spite of Gower's clear description of the different types of involuntary movement observed, and the fact that those which occurred in many of the children were more like chorea or even dystonia than true athetosis, this term continued to be used to describe the category.

Not all authors distinguished cases of "Athetosis" from those suffering from generalised rigidity or bilateral hemiplegia. They pointed out that involuntary movements, especially true athetosis, occurred not infrequently in patients with generalised rigidity and that transitional pictures between "Athetosis" and "Generalised Rigidity" had been described.

The work of Freud marked an advance in the classification of cerebral palsy. His classification was based on clinical findings, though he hoped that further study would eventually differentiate disease entities which were aetiologically, pathologically and clinically distinct within his categories.

He /

He distinguished between unilateral and bilateral impairment of motor function. The latter he termed "The Cerebral Diplegias". His classification was as follows -

Hemiplegia
Cerebral Diplegia, comprising - Generalised rigidity
Paraplegic rigidity
Bilateral Hemiplegia
Generalised chorea and double athetosis.

He emphasised the similarity in aetiological and clinical findings of patients with generalised rigidity and paraplegic rigidity and considered them to differ in degree of severity of their clinical manifestations rather than in kind. They were characterised by more or less symmetrical paresis of the limbs more marked in the lower limbs than the upper. There was often associated strabismus, mental defect and epilepsy. A high proportion of patients were prematurely born and a history of birth injury was frequent. Generalised and paraplegic rigidity could be distinguished from bilateral hemiplegia, because in the latter the paresis was more severe in the upper limbs than the lower and pseudobulbar palsy was constant and more severe than was encountered even in cases of severe generalised rigidity. Severe mental defect and epilepsy were almost always associated with bilateral hemiplegia.

Freud recognised that other categories of cerebral palsy might be required, and in particular suggested that congenital, non-progressive ataxia, similar in other respects to /

to familial spinocerebellar ataxia, might eventually form one.

Aetiological Classification. The very title of Little's second paper suggests an aetiological definition of cerebral palsy - "On the influence of abnormal parturition, difficult labour, premature birth and asphyxia neonatorum on the mental and physical condition of the child, especially in relation to deformities". Yet Little was aware of the fact that clinical syndromes could result from postnatal disease or injury of the nervous system which were similar to those which resulted from birth injury. In particular, he noted that hemiplegia could follow fevers and convulsions in early childhood. The early French authors also distinguished between congenital and acquired hemiplegia, the latter most commonly occurring as a complication of infectious diseases in the first three years of life. (Cazauvielh, 1827; Cotard, 1868; Cruvehlier, 1862). The clinical course of "Acute infantile hemiplegia" was studied in great detail by Wuillamier, (1882).

Diplegic syndromes were much less often acquired in postnatal life than hemiplegia, but Freud presented a number of cases of acquired paraplegia, generalised rigidity and generalised chorea. (Freud, 1897). He also described a number of hereditary or familial forms of cerebral palsy which he considered separately from those apparently resulting from birth injury. He examined the possibility of classifying cerebral palsy in aetiological categories, but came to the conclusion that in the contemporary state of knowledge this /

TABLE I.

The aetiological classification of cerebral palsy suggested by Sachs 1891

Groups	Clinical forms in order of frequency.
Paralysis of intra uterine onset.	Diplegia Paraplegia Hemiplegia
Birth palsies	Diplegia Paraplegia Hemiplegia Diataxia Cerebellar form
Acute (acquired) palsies	Hemiplegia Paraplegia Diplegia Choreic and athetoid disorders, unilateral and bilateral

this was impossible. Too little was known about the pre-natal environment of patients and it seemed apparent that very similar clinical pictures could be caused by different aetiological factors. Frequently, also there were multiple aetiological factors.

Nevertheless, attempts to classify by aetiology were made. The most detailed scheme was that of Sachs, (1891). (Table I).

It will be observed that Table I contains categories which are not mutually exclusive and that the objections to aetiological classification expressed by Freud appear to be well founded. Sachs himself later abandoned his aetiological classification in practice, and returned to classification on the basis of neurological signs. (Sachs and Hausman, 1926).

Modern Classifications of Cerebral Palsy. The increase of philanthropic and therapeutic interest in cerebral palsy in the early nineteen thirties meant that many clinicians and therapists with differing outlooks became concerned with the diagnosis and treatment of the condition. A number of classifications of cerebral palsy have been produced since this time which reflect these differences in attitude; the academic interest of the neurologist, the more practical points of view of orthopaedic surgeons and therapists, the conflicted viewpoints of the paediatrician.

The classifications most widely used in the past twenty years /

years are those based on the work of Phelps, (1940, 1941, 1943, 1950). Though these have varied in detail from year to year their basis has remained the same. Phelps abandoned classification on the basis of complete neurological syndromes and substituted for it classification based primarily on the state of muscle tone and the presence or absence of involuntary movement. Further classification takes account of the number of limbs which are abnormal, aetiological factors, the presumed site of neuropathological changes and associated defects of the senses. (Table 2).

The aim of this classification and others like it is to give brief descriptions of clinical manifestations which are likely to be helpful to therapists and others concerned with the practical management of patients. To some extent it succeeds in this. For example the detailed sub-categories of "Athetosis" allow almost any patients with involuntary movement to be classified in one or another of them. But in practice differences in the classifications of such patients between different clinicians, and even by the same clinician at different times, are frequent. The sub-categories are not mutually exclusive. For example, patients with involuntary rotatory movements of the limbs frequently show dystonic movements also.

Major criticisms of this scheme of classification are that muscle tone varies greatly in patients as they mature, and as the fully established picture of their cerebral palsy gradually develops. Patients with "Little's disease", for example, /

TABLE 2.

Recent American Classification of Cases of Cerebral Palsy based on that of Phelps by Hellebrendt 1950-51

I. Spastic.
- A. Aspastic.
- B. Spastic.
  - (1) Monoplegia.
  - (2) Hemiplegia.
  - (3) Paraplegia.
  - (4) Triplegia.
  - (5) Quadriplegia.
- C. Basilar.

II. Athetosis.
- A. Tension.
- B. Non-Tension.
- C. Dystonic.
- D. Flail.
- E. Arm Neck.
- F. Deaf.
- G. Shudder.
- H. Hemi-Athetoid.
- I. Cerebellar Release.
- J. Rotary.
- K. Emotional Release.
- L. Tremor.
- M. Unclassified.
  - (1) Paraplegia.
  - (2) Quadriplegia.
  - (3) Monoplegia.
  - (4) Recovered.

III. Rigidity.
- A. Intermittent.
- B. Continuous.
- C. Miscellaneous.
  - (1) Hemiplegia.
  - (2) Paraplegia.
  - (3) Triplegia.
  - (4) Quadriplegia.

IV. Tremor.
- A. Intention.
- B. Constant.

V. Ataxia.
- A. Cerebellar.
- B. 8th. Nerve

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- C. Basilar.

II. Athetosis.
- A. Tension.
- B. Non-Tension.
- C. Dystonic.
- D. Flail.
- E. Arm Neck.
- F. Deaf.
- G. Shudder.
- H. Hemi-Athetoid.
- I. Cerebellar Release.
- J. Rotary.
- K. Emotional Release.
- L. Tremor.
- M. Unclassified.
  - (1) Paraplegia.
  - (2) Quadriplegia.
  - (3) Monoplegia.
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- A. Intermittent.
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- B. 8th. Nerve

example, are commonly hypotonic for some weeks after birth, pass through a stage when dystonic movements of the trunk are the major clinical finding, may show a stage when anti-gravity hypertonus, (rigidity) is preponderant, and only after weeks or months is true flexor hypertonus, (spasticity) apparent. Using Phelps terminology, repeated re-classification of these cases is necessary.

Moreover, the categories are not well defined. There are many causes of stiffness of the limbs which appears to be all that "Rigidity" implies. In this category, for example, may be found cases of hemiplegia with stiffness due to contracture, and cases of diplegia with stiffness due to antigravity hypertonus.

There is much nosological confusion in the definition of the sub-categories. Under "Spastic", for example, are found sub-categories of "Aspastic", "Spastic", and "Basilar". These are not mutually exclusive, and in any case, "Aspastic Spastic" is not a very meaningful description.

A large number of classifications are essentially modifications of that suggested by Phelps, for example, those of Asher and Schonell, (1950), Andersen, (1954), Collis et al, (1956). In all of them, muscle tone and the presence or absence of involuntary movement are the major initial criteria of classification, though some lay more stress on topographical classification than does Phelps. Thus, Collis et al, distinguish a category of hemiplegia from other forms of "Spastic paralysis". (Collis et al, 1956).

Attempts /

Attempts to simplify the classification of cerebral palsy have been made, largely by paediatricians. The majority of these attempt to classify on the basis of muscle tone and the presence or absence of involuntary movement, together with topographical distribution of the motor defects. (Evans, 1948; Wyllie, 1951). Table 3). In practice, these classifications result in fairly adequate descriptions of the common motor defects encountered. Thus it is possible to refer to spastic hemiplegia and triplegic athetosis. But it is difficult to distinguish between tetraplegic patients whose greatest neurological involvement is in the lower limbs from those with bilateral hemiplegia. Yet this distinction is important enough to lead Evans to make an exception for it in his system of classification. Moreover, classifications of this type do not give any indication of the stage of development of the clinical syndrome described, nor of its severity.

Attempts to overcome these defects have been made by describing each category of cerebral palsy in a number of different terms. The most comprehensive classification to do this is that of Perlstein, (1952, 1955). He suggests a classification based on presumed site of pathology, clinical manifestations, topographical description, severity of involvement, muscle tone and aetiology. (Table 4). The authors claim that this permits the accurate description of almost every case of cerebral palsy seems justified, but the distinctions between the categories are not all valid.

For /

TABLE 3.

Classification of Wyllie, 1951

I. Congenital symmetrical diplegia.

2. Congenital paraplegia.

3. Quadriplegia or bilateral hemiplegia.

4. Triplegia.

5. Hemiplegia with the additional qualifications -

   (a) spasticity
   (b) flaccidity
   (c) mixed types
   (d) athetosis
   (e) ataxy

TABLE 4.

Classification of Perlstein, 1952

By Clinical Symptoms	Topographical Involvement of extremities	By Muscle Tone	Severity	Etiology
Spastic conditions	Paraplegia	Isotonic	Mild	A. Prenatal
Dyskinesias	Diplegia	Hypertonic	Moderate	1. Hereditary
Choreas	Quadriplegia or Tetraplegia	Hypotonic	Severe	(a) Static
Athetoids	Hemiplegia			(b) Progressive
Dystonia	Triplegia			2. Acquired in Utero
Tremors	Monoplegia			(a) Infection
Rigidity	Double Hemiplegia			(b) Anoxia
Ataxia	Limited to both upper extremities			(c) Cerebral haemorrhage
				(d) Rh. factor
				(e) Metabolic disturbance
				(f) Gonadal irradiation
				B. Natal Factors
				1. Anoxia
				2. Cerebral haemorrhage
				(a) Trauma
				(b) Pressure change etc.
				C. Postnatal Factors
				1. Trauma
				2. Infections
				3. Toxic causes
				4. Vascular accident
				5. Anoxia
				6. Neoplasms and develop-

TABLE 5.

CLASSIFICATION OF CEREBRAL PALSY*
(Minear 1956)

I. Physiological (motor)

A. Spastic

B. Athetotic

1. Tension
2. Non-tension
3. Dystonic
4. Tremor

Types of athetosis acceptable by the American Academy for Cerebral Palsy to date. Four outline of [illegible] of athetosis described by Wi[illegible] Phelps see "Types of Cerebral [illegible] Defined" herein.

C. Rigidity

D. Ataxic

E. Tremor

F. Atonic (rare)

G. Mixed

H. Unclassified

II. Topographical

A. Monoplegia
Involves one limb; condition is rare; should be checked closely to determine if you are not dealing with a para[illegible]a or hemiplegia.

B. Paraplegia
Involves the legs only and practically alw[illegible] the spastic or rigidity type.

C. Hemiplegia
The lateralized one-half of the body is af[illegible] and it is usually spastic, although pure athetoid hemiplegia[illegible] ccasionally seen, as are pure rigidity hemiplegias. There [illegible] n sensory involvement in the areas of proprioception to poi[illegible] rimination and form perception. Aphasias appear more fr[illegible] y in right than in left hemiplegias and are much more comm[illegible] he acquired than in the congenital cerebral palsy.

D. Triplegia
Involves 3 extremities, usually both leg[illegible] arm, usually spastic. This may represent hemiplegia plus para[illegible] incomplete quadriplegia. In the latter case, both arms will[illegible] ual or nearly equal in length. In the former, the involved ar[illegible] be shorter.

E. Quadriplegia
(Tetraplegia) Involvement of all 4 extremi[illegible] Patients with the greatest involvement of the legs are u[illegible] spastic, and patients with greatest involvement of the [illegible] e usually the dyskinetics, including athetoids.

F. Diplegia

This term is seldom used. "Paralysis affe[illegible] like parts on either side of the body; bilateral paraly[illegible] (The American [illegible] Medical Dictionary, Dorland, 21[illegible] ition).

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G. Double Hemiplegia

This term is seldom used. " ..... implies those cases in which the arms are more involved than the legs. These are usually spastic in type". (Cerebral Palsy - Its Individual and Community Problems, edited by William M. Cruickshank, Ph.D. and George M. Raus, M.D., Syracuse University Press, 1955).

III. Etiological (abbreviated)

A. Prenatal

B. Natal (abbreviated)

C. Postnatal

IV. Supplemental

A. Psychological evaluation

(1) Degree of mental deficiency, if any.

B. Physical status

(1) Physical growth evaluation (Wetzel Grid or other)
(2) Developmental level (Gesell)
(3) Bone age
(4) Contractures

C. Convulsive seizures

D. Posture and locomotive behaviour patterns

E. Eye-hand behaviour patterns (abbreviated)

F. Visual status

(1) Sensory
(a) Amblyopia
(b) Field defects

(2) Motor

G. Auditory status
(1) Pitch range loss
(2) Decibel loss

H. Speech disturbances.

V. Neuroanatomical (See sub-headings under "Brain", topographic headings, Standard Nomenclature of Diseases and operations).

Failure to discuss the parts of the brain involved in the different motor types of cerebral palsy is deliberate. This discussion is reserved until after we correlate the lesions from a sufficient number of brains from patients with cerebral palsy with the symptoms to gain a scientific understanding of the problem.

The following two headings are added for the sake of completeness, and for the use of the clinician studying the cerebral palsied patient. They are not intended to be used for coding by the medical record librarian:

VI. Functional Capacity (degree of severity)

Class I. Patients with cerebral palsy with no practical limitation of activity.
II. Patients with cerebral palsy with slight to moderate limitation of activity.
III. Patients with cerebral palsy with moderate to great limitation of activity.
IV. Patients with cerebral palsy unable to carry on any useful physical activity.

VII. Therapeutic

Class A. Patients with cerebral palsy not requiring treatment.
B. Patients with cerebral palsy who need minimal bracing and minimal therapy.
C. Patients with cerebral palsy who need bracing and apparatus, and the services of a cerebral palsy treatment team.
D. Patients with cerebral palsy limited to such a degree that they require long term institutionalization and treatment.

For example, he distinguished under "Topographical", patients with "quadriplegia" and "bilateral hemiplegia". There is no suitable category for diplegic patients with tetraplegic rigidity. These are not major criticisms. The main objection to the classification is that its informality encourages different interpretations by different clinicians. Series of cases cannot easily be compared when this classification is used.

A rather similar classification in which categories are defined in a number of different ways was devised by The American Academy for Cerebral Palsy. (Minear, 1956). (Table 5.). This was produced after the members of the Academy had been asked what basis of classification they preferred, and comparing the fifty-five replies received. This procedure might be compared to circulating the Bishops in the Church of England about the catechism before deciding upon its revision.

The major criteria for classification are clinical findings. Muscle tone and the presence or absence of involuntary movements were the most important of these and the major categories of Phelps were retained. Further classification was by distribution of the motor abnormalities, aetiology and supplemental findings. Indications of the neuroantomical basis of the cerebral palsy, the functional capacity of the patient and his therapeutic possibilities should also be given when possible. The major objections

to /

to this classification is that formal distinctions between the categories under the heading of "Topographical", are not valid. In particular, Diplegia and Double Hemiplegia are descriptions of neurological syndromes, not of topographical distribution. They are not of the same order as Monoplegia, Paraplegia hemiplegia, Triplegia and Quadriplegia. The factors given under "III Etiological" and "IV Supplemental" are commonly found in combination and do not form a satisfactory basis for classification, though they may be helpful in clinical description.

Classification by neurological syndromes has continued to be made by some authors. A number of classifications by neurological syndromes attempt to break down the major categories described by Freud, by taking out of them the rarer hereditary and familial forms of cerebral palsy with characteristic aetiological, clinical and pathological findings which have been recognised since his time. They also attempt to differentiate further the clinical syndromes on pathological or prognostic grounds. A typical example of this type of classification is that of Wilson, (1940). (Table 6.). It will be observed that it is rather complex and the tendency to mix aetiological and clinical criteria makes for nosological confusion and overlapping of categories. Thus, it is hardly suitable for practical use.

Other neurologists are sufficiently impressed by the diversity of the conditions which comprise cerebral palsy that they do not attempt formal classification under this heading. /

TABLE 6

Classification of patients suffering from Cerebral Palsy by Wilson (1940)

(The author does not make the distinction between cerebral palsy on the one hand, and degenerative conditions of the nervous system which is current today).

I. Little's disease (prenatal or natal)

(a) Obstetrical cases.
(b) Haemorrhagic disease of the newborn.
(c) Intrauterine infection.
(d) Primary agenesis of the pyramidal system.
(e) "etat marbré" of the corpus striatum.

2. Cerebral diplegia with or without involuntary movements, epilepsy and idiocy (restricted to encephalopathies of postnatal origin).

(a) Simple diplegia. Limited.
Generalised.
(b) Diplegia with involuntary movements; double athetosis.
(c) Pseudobulbar diplegia (with and without athetosis)
(often found in association with general diplegia)
(d) Progressive and familial diplegia.

3. Infantile cerebral hemiplegia.

4. Cerebellar diplegia.

(a) diplegia with cerebellar symptoms. Cerebrocerebellar diplegia.
(b) atonic diplegia. (atonic-astatic type of Foerster, 1910)
(c) cerebellar diplegia. (Congenital cerebellar ataxia of Batten 1950)

5. Hereditary (familial) spastic paraplegia or diplegia.

TABLE 7

Classification of Benda, 1952

1. Little's spastic rigidity (decerebrate rigidity).
2. The pyramidal type (monoplegias, hemiplegias, diplegias, paraplegias).
3. The mixed extra-pyramidal-pyramidal type (paraplegias with athetosis).
4. The ataxic-atonic cerebellar type.

TABLE 8

Classification of Courville, 1954

A. Cortical syndromes.

   1. Spastic diplegia-paralysis mainly in the lower limbs; the upper limbs may retain a relatively normal motor function.
   2. Hemiplegia.
   3. Double hemiplegia.
   4. Monoplegia.

B. Ganglionic syndromes.

   Symptoms referable to the basal ganglia which may constitute the major manifestations of the clinical picture or be only one element in it.

C. Cerebellar syndromes.

heading. The arbitrary nature of the definition of "Cerebral Palsy" means that there is much to be said for considering the conditions which it comprises in the more general context of nervous disease in childhood. (Ford,1952). On the other hand, in practice there are a number of non-progressive diseases of the nervous system in childhood which present certain common clinical and therapeutic problems. This justifies some attempt to define and classify them.

A strong objection to many of the classifications on the basis of neurological syndromes is that they are not expressed in clinical terms. Thus, the term "Pyramidal forms" is used quite commonly to define a category containing hemiplegia, and symmetrical paresis of the limbs not associated with involuntary movement. (Colli Grisoni, 1955; Benda, 1957). Table 7). Similarly, though his classification is essentially by neurological syndromes, Courville divides his categories into those which originate from disorders of the cortex, those due to abnormalities of the basal ganglia and those due to cerebeller lesions. (Courville, 1954). (Table 8). Clearly clinical classification should be on the basis of observed clinical findings, not on the basis of the presumed neuropathology. Another weakness of most recent classifications by neurological syndromes is that they do not take account of complicated or "mixed" forms of cerebral palsy. For example, patients with hydrocephalus often show ataxia and paresis more marked /

marked in the lower limbs than the upper, associated with spastic increase of tone in the limbs and extensor plantar responses. Such patients could be placed in at least two categories in all the three classifications quoted above.

The Present Classification. The classification of cerebral palsy used in this thesis is primarily by neurological syndromes. It is based on that of Freud with modifications made necessary by advances in knowledge since his time. (Ingram, 1955; Balf and Ingram, 1956).(Table 9)

Two new categories of cerebral palsy are introduced. These are "Ataxia" and "Ataxic Diplegia". Freud recognised that clinical syndromes of congenital ataxia might become well enough defined to merit separate categories being made for them. Within ten years of his writing this, Batten had defined at least two ataxic syndromes in clinical terms. (Batten, 1903, 1905). The first of these consisted of cerebellar ataxia alone, with weakness, incoordination and hypotonia in all four limbs. The second which was rarer comprised cerebellar ataxia with paresis in the limbs, spastic increase of tone and extensor plantar responses. The first condition is called "Ataxia", the second "Ataxic Diplegia" in this classification. Some recent series of children with cerebral palsy have contained five per cent. or more of patients with ataxia as their predominant motor defect. (Hellebrandt, 1950-51; Skatvedt, 1958; Andersen, 1954; Woods, 1957). There is need to modify Freud's classification in order that they can be placed in distinctive /

TABLE 9

Classification of cases of cerebral palsy in childhood
(Ingram 1955; Balf and Ingram 1955)

Neurological diagnosis	Extent	Severity
Hemiplegia	(Right (Left	(Mild (Moderately severe (Severe
Bilateral hemiplegia		(Mild (Moderately severe (Severe
Diplegia		
Hypotonic) Dystonic) Rigid or spastic)	(Paraplegic (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Ataxic diplegia		
Hypotonic) Spastic)	(Paraplegic (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Ataxia		
Cerebellar) Vestibular)	(Predominantly (Unilateral (Bilateral	(Mild (Moderately severe (Severe
Dyskinesia		
Dystonic) Choreoid) Athetoid) Tension) Tremor)	(Monoplegic (Hemiplegic (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Other		

distinctive categories.

Some re-arrangement is made in Freud's classification of patients suffering from bilateral cerebral palsy which he termed "Diplegia". (Freud, 1893, 1897). Cerebral palsy in which involuntary movements are the major motor defect is made a major category. "Dyskinsia"; this term suggested by Perlstein, appears to be more comprehensive and less limiting than the "Bilateral athetosis and generalised chorea" of Freud. (Perlstein, 1952). Freud himself suggested that as more came to be known about them, syndromes characterised by involuntary movements might well require to be placed in a category distinct from "Generalised rigidity" and the other forms of "Diplegia". Since between 10 and 30 per cent. of children with cerebral palsy in most series are considered to show involuntary movements as their major clinical feature, there is practical as well as theoretical justification for making a separate major category for them.

Freud distinguished between "Bilateral Hemiplegia" and "Generalised Rigidity" on clinical grounds. In bilateral hemiplegia paresis was more marked in the upper limbs than the lower; there was more or less severe involvement of the bulbar musculature, mental defect and epilepsy were almost constant. In "Generalised rigidity" increased tone of the limbs was often more striking than paresis. The lower limbs were always more affected than the upper and pseudo-bulbar palsy was rarely severe. Mental defect and epilepsy were less constant. His distinction between the conditions has stood /

stood the test of time and it seems justifiable to make a major category for "Bilateral Hemiplegia".

The term "Diplegia" is then left to apply only to Freud's categories of "Generalised Rigidity" and "Paraplegic Rigidity". He himself emphasised the clinical similarity between these conditions and seems to have regarded them as different in degree rather than in kind. It is easy to make a simple common definition for them and they are placed in a single category "Diplegia" in the present classification. (Courville, 1954).

A category of "Other" is provided for the few cases which cannot be classified in the other defined categories. Most of the patients included in this group show clinical features characteristic of more than one of the other categories. A few have clinical findings which are not included in them.

The basis of further classification.

The clinical picture of cerebral palsy commonly changes as the child matures. These changes are usually gradual, but it is possible to define arbitrarily stages in the changing clinical picture which may be recognised. Thus, diplegic patients often show "rigidity" of the limbs with predominant extensor positions of the limbs and trunk and an increase in antigravity tonus for a period of months before become "spastic", with relative increase in flexor tonus of the limbs and trunk and a tendency to positions of flexion and flexor contracture. Each stage has its characteristic patterns /

patterns of motor behaviour or levels of maturation of motor function. For example, prehension is never seen in the "rigid" stage of diplegia, though it may be observed in many patients in the "spastic" stage. Much information of value may be conveyed by indicating the stage of development of the cerebral palsy in classification.

Further classification takes account of the impairment of motor function which the cerebral palsy has caused. The number of limbs significantly affected is indicated. Minor neurological abnormalities in the limbs which do not result in any detectable loss of function in everyday life are ignored. They are indicated by the neurological diagnosis. The severity of the cerebral palsy is gauged by the reduction of activities in everyday life caused by the condition in comparison with normal children of the same age. The activities of which account is taken are different in the different categories. For example, the severity of the handicap in hemiplegia is measured by the residual function of the affected upper limbs, and in diplegia by the use the child has of the legs.

Account is not taken in the classification of defects of the special senses, mental impairment, the presence or absence of epilepsy, sensory loss in the limbs or secondary skeletal abnormalities. They they are important these can be indicated in the description of patients, but their routine use would overburden the classification.

For /

For the same reasons, no attempt to classify by aetiological factors is made formally. These are known with certainty in only a minority of patients. They may easily be indicated in them by adding brief description. For example, a proportion of patients in the category of "Dyskinesia" have suffered from kernicterus, and some of those with ataxic diplegia show hydrocephalus. Examples might be "Moderately severe tetraplegic dyskinesia showing choreoathetosis and dystonia following kernicterus" or "Severe spastic ataxic diplegia with tetraplegic involvement associated with hydrocephalus".

The Clinical Categories.

Hemiplegia. Hemiplegia is a unilateral paresis, usually associated with some spasticity and flexion contracture of the limbs. The upper limb is nearly always more severely affected than the lower. In most congenital cases, and in those arising in early childhood, growth is retarded in the affected limbs so that these are shorter and thinner than those on the unaffected side. Vasomotor disturbances may occur, but their severity is not necessarily proportional to the severity of the paresis or to the degree of dwarfing. Athetosis of the fingers and toes is a frequent finding, but since this is clearly a complication of the hemiplegia, and not the major disability, cases showing it are not classified as suffering from dyskinesia. Sensory disturbances in the affected limbs are common; an associated hemianopia may be found.

In /

In classification the side affected should be noted as the disability tends to be greater if the master hand is involved, and speech disorders are commoner when this is so. Severity is assessed by residual function. Mild cases can use the affected hand independently for everyday activities. Moderately severely handicapped patients use the hand only as an assistant to the other. In severely affected cases there is no useful function in the hemiplegic hand, though the limb may fulfil a supporting role when what would normally be bimanual activities are attempted.

Bilateral Hemiplegia. These patients show tetraplegic paralysis greater in the upper limbs than the lower, usually with moderate or severe flexion contracture and spastic increase of tone. There is frequently severe dysarthria, dysphagia and a liability to descending respiratory infections due to pseudobulbar paresis. Most patients are mentally defective, epileptic, and severely microcephalic. Developmental malformations are commonly associated.

It is necessary to distinguish this group of patients from those with hemiplegia and diplegia. The total disability exceeds the summation of two hemiplegias in the severity of the functional motor impairment of the limbs and the involvement of the bulbar musculature. The greater motor impairment in the upper limbs, the presence of more or less severe bulbar palsy and the absence of the characteristic "stages" of development found in diplegia differentiate from this condition.

All /

All cases are severely disabled. Mildly affected patients retain some use of the upper limbs, moderately severe cases have some use of the lower limbs, and those who are severely handicapped retain no use of their limbs being bedridden and helpless.

Cerebral Diplegia. Though not well derived, the term cerebral diplegia, or diplegia, is useful when employed to describe patients with more or less symmetrical paralysis worse in the lower limbs than the upper, and dating from birth or shortly afterwards. (Freud, 1897; Evans, 1948; Courville, 1948; Ingram, 1955b). Fine movements of the fingers and toes are invariably impaired. There is usually a marked increase of muscle tone and relative dwarfing below the waist. Mental impairment, epilepsy and strabismus are common. Bulbar paresis is rarely severe. Approximately 40% of patients are prematurely born and the majority have a history of perinatal hypoxia.

The clinical findings in diplegia alter as the child matures. (Ingram, 1955b). A number of arbitrarily defined stages may be recognised in a high proportion of patients. The duration of these stages is very variable in different cases, and in the most severely affected patients the progression through stages may stop short of the development of the final picture.

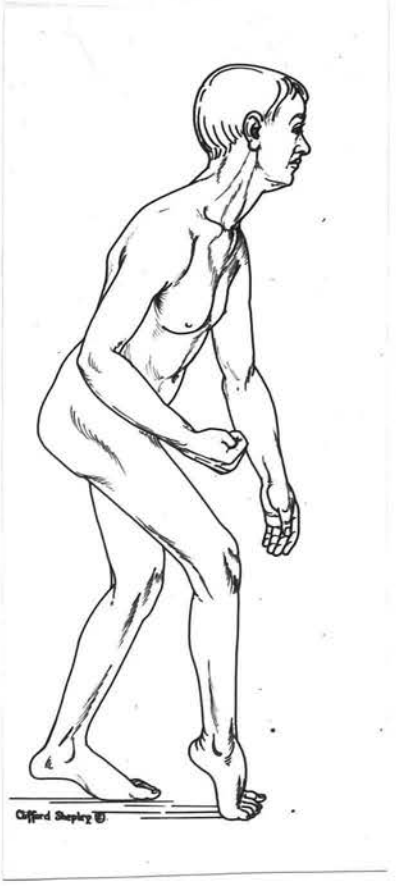
In the first few weeks of life the child is often thought to be normal, but careful observation will reveal a marked poverty of movement and generalised hypotonia. In severely affected /

affected patients these abnormalities are usually observed by the parents, at least in retrospect, though commonly ascribed to the effects of prematurity or the result of "laziness" by the medical attendant. This may be termed the "hypotonic" stage of diplegia. After weeks or months the next stage begins. It is characterised by the occurrence of sudden hypertension of the neck, back and limbs when the child is alarmed or handled. The position assumed by the child is essentially one of opisthotonos. The movement which produces this position may be described as being dystonic. It may be produced most easily during clinical examination by extending the child's head suddenly. A position of opisthotonos with generalised rigidity of the trunk and limbs results and persists for a few seconds, although at rest with the head in any other position, the muscles will be found to be hypotonic. This may be termed the "dystonic" stage of diplegia. (Fig. I.)

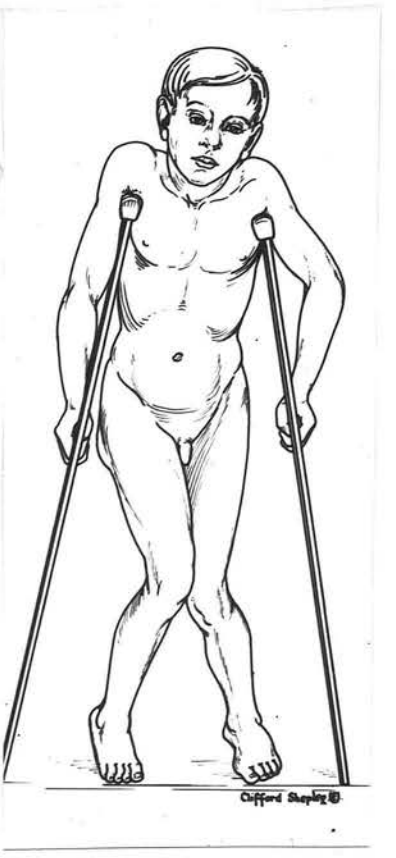
Very severely affected patients suffering from diplegia may remain in the "hypotonic" or "Dystonic" stages of the condition for the rest of their lives, but the majority of cases show some maturation of neuromuscular function. Their clinical findings change as maturation occurs.

After a variable period the stage of dystonia is succeeded by the development of rigidity in the majority of patients. At first, a persistent stiffness of the limbs is noted at rest and they tend to be held in extended positions. /

Postures in the different stages of diplegia.(Continued)



d.



e.

Late rigid/spastic diplegia showing flexion contractures of elbows,hips and knees.The increase of tone is predominantly of spastic type when these postures occur.

positions. The stiffness is due to a relative increase in the tone of antigravity muscles ("extensor spasticity"). Although postural reflexes and the symmetrical and tonic neck reflexes are usually very easily elicited during this stage, the tendon reflexes are not usually increased. At this stage of diplegia the clinical state might be described as one of generalised rigidity without exaggeration of the tendon jerks. A proportion of patients will always show this type of diplegia, which may be interpreted as an arrested state of development of the condition. In the majority of patients, however, a spastic increase of tone (flexor spasticity) gradually appears and as this increases, antigravity hypotonus diminishes. The resting position becomes one of generalised flexion, or semiflexion; the tonic and symmetrical neck reflexes become much less marked. The biceps, triceps, supinator, knee and ankle jerks become exaggerated. The plantar responses which have previously been infantile or equivocal become more definitely extensor. There is a tendency to contractures in flexion. This is the final stage of diplegia and the only form of the condition which is adequately described in textbooks. (Ford, 1952). (Fig.I)

Since the stages in the development of diplegia appear to be well defined and easily differentiated it is desirable to note them in classification. It is also necessary to classify patients according to the severity and extent of their functional impairment in everyday life. They are, therefore, placed in groups of paraplegia, triplegia and tetraplegia. /

tetraplegia. Severity is best assessed on the basis of efficiency in walking, since it is the lower limbs which show the major disability. Mild cases are those whose gait is clumsy rather than disabled; moderately severe cases are those with an obviously impaired gait who cannot run and are usually somewhat unsteady when walking. Severe cases are those who can walk only with some support well after the age at which independent walking might be expected in a normal child.

Ataxic Diplegia. In this condition there is some weakness and incoordination of voluntary movement, commonly associated with intention tremor and inability to perform fine or rapid movements well in all four limbs. There is unsteadiness and a tendency to walk on a broad base when in the erect position. In addition, there is paresis of voluntary movement, more severe in the lower limbs than the upper commonly associated with spastic increase of tone. Adductor spasm is usually absent and is never a conspicuous feature. The plantar responses are extensor.

The ataxia appears to be of cerebellar type clinically, but because of the severe paresis it is frequently impossible to assess its importance in causing functional motor impairment. The patients are commonly hypotonic during the first weeks or months of life, but they never show dystonic or rigid stages. This, together with slight degree of adductor spasm differentiates it from diplegia. The presence of bilateral paresis with spasticity, more marked in the lower limbs /

limbs distinguishes ataxic diplegia from uncomplicated ataxia.

A family history of neurological disorders such as epilepsy, mental defect, or ataxic diplegia is often present. A number of patients show hydrocephalus of congenital origin or acquired as a result of meningitis or in early childhood.

Mild cases are those without significant functional loss, though they are clearly affected and recognisably clumsy. Moderately severe cases have some limitation of normal activities. Severe cases are those who need to be helped in all but the simplest everyday activities. It is useful to indicate whether the muscles in the lower limbs are hypotonic as they are in the most severely affected patients, or hypotonic as they are in those with a more fully developed and matured clinical picture.

Ataxia. Patients in this category show weakness, incoordination of movement usually associated with intention tremor, and impaired balance as their presenting features. The ataxia may be symmetrical or chiefly unilateral. Mild cases have no significant loss of function in everyday activities, though they are recognised to be abnormally clumsy. Moderately severe cases suffer from some limitation of normal activity, but can walk independently with or without support. Severely affected patients require help in all but the simplest everyday activities and cannot walk independently with or without support at an age much older than that at which a normal child acquires independent walking balance. /

balance.

The great majority of patients suffer from ataxia of cerebellar type. A proportion of them suffer from developmental abnormalities of the cerebellum, but in others the symptoms appear to be the result of birth injury.

Dyskinesia. In this category involuntary movements of the limbs produce the major motor disability. (Perlstein, 1952).

Involuntary movements may occur in a number of other forms of cerebral palsy as a complication. Athetosis, the slow writhing movements of the fingers predominantly involving abduction and extension, described by Hammond, commonly complicates hemiplegia in childhood. Rarely choreoid or even dystonic movements may occur in hemiplegia. (Gowers, 1876). Dystonic movements are characteristic of an early stage of diplegia, and later in the spastic stage athetosis may be found in a minority of patients. Occasionally athetosis may also be observed in patients with ataxic diplegia. In these conditions, however, involuntary movements are not the major cause of motor disability as they are in dyskinesia.

The types of involuntary movement which occur in dyskinesia are classified as suggested by Perlstein, (1952). Dystonic movements are those which involve predominantly the proximal parts of the limbs and the trunk and are slow and writhing in quality. They commonly tend to place the parts of /

of the body which they affect in the "opisthotonic position". Choreoid movements also affect the proximal parts of the limbs, but they are more rapid than those classified as being dystonic. limbs often appear to have been thrown out of position by them. They may also affect the trunk and face, resulting in sudden erratic changes in posture, respiratory rhythm and in grimacing.

Athetoid movements affect the fingers and toes predominantly, though in severe cases the wrists, forearms and occasionally even the elbows may also be affected. The movements are slow and writhing. They are accentuated by voluntary activity, but are absent at rest. They are often best seen in the form of extension and abduction of the fingers and thumb when the child reaches.

Sudden involuntary variations of muscle tone, commonly a sudden generalised hypertonus termed "tension", affecting both flexor and extensor muscle groups also occur commonly in dyskinesia. The whole limb becomes stiff during attempted voluntary movement and bizarre postures may occur transiently as a result.

Tremor is a regularly occurring involuntary movement, which is commonly, though not invariably made more obvious when voluntary movement is attempted. Tremor is rarely the major cause of motor disability.

More than one form of involuntary movement is commonly found in patients suffering from dyskinesia. Thus, in patients who have suffered from kernicterus, the result of rhesus incompatability, there is commonly dystonia, choreoid movements and athetosis athetosis /

athetosis, associated in the majority with "tension", the sudden involuntary generalised increase of muscle tone which has been described. The types of involuntary movement observed may be indicated when the patient is classified, though it must be admitted that descriptions such as "a dystonic choreoathetoid with tension" are large mouthfuls.

As in diplegia a number of arbitrarily defined stages of development may be recognised in dyskinesia. After the complications of the neonatal period have been survived, patients are commonly somewhat immobile and hypotonic for some weeks. After this time the same tendency to assume positions of opisthotonos as was noted in diplegia, may be observed. As neuromuscular maturation proceeds this dystonic movement becomes less generalised, affecting the trunk to a diminishing extent and being confined progressively to the limbs. As an increased amount of voluntary activity becomes possible in the limbs, the involuntary movements of the proximal parts become more rapid and are more accurately described as choreoid though they rarely reach the speed of those observed in rheumatic chorea. When neuromuscular maturation reaches the stage of allowing voluntary movements of the hands, athetoid movements may also be observed. Thus, the description of the involuntary movements observed in patients also gives some indication of the stage of dyskinesia from which they suffer. Children whose movements are predominantly dystonic show a lower

stage /

stage of neuromuscular development than those whose movements are predominantly choreoid. They have less voluntary control of the posture of their heads and trunks and the voluntary movement patterns of their limbs are less mature. For example, voluntary supination of the forearms is invariably impossible.

Further classification of children suffering from dyskinesia takes account of the number of limbs which are functionally impaired by involuntary movements in everyday life and the severity of the handicap. The latter is gauged using the same criteria as were applied with ataxic patients.

Other categories. Few patients suffering from cerebral palsy cannot be placed in one of the categories which have been described.

Occasional patients do show features of more than one category. Some authors consider that 5 or 10% of patients suffer from "Mixed Cerebral Palsy" and a few even make a special category into which they may be classified. (Asher and Schonell, 1950; Hellebrandt, 1950).

The descriptions given of many such "Mixed Cases" suggest that the patients are, in fact, suffering from diplegia or dyskinesia in their early stages. Since the clinical features of the early stages do not appear too widely known they are regarded as comprising "Atypical" or "Mixed forms". There is difficulty in classifying a proportion of patients, however, /

however, especially those under the age of one year, and they may be placed in this category until accurate diagnosis becomes possible.

A few patients do show forms of cerebral palsy which are not readily classified in the categories which have been defined, and these may conveniently be classified as suffering from "other types". They include children with post-encephalitic syndromes in which ataxia and involuntary movement are prominent. Patients in whom cerebral palsy is a complication of other medical conditions, for example, these with spastic paresis associated with arthrogryposis multiplex may also be conveniently placed in this category.

Since the term "other types" comprises such a diversity of neurological conditions, it is necessary that these should be described as accurately as possible when they are classified.

Discussion of the Present Classification.

The classification had its origin in the need to classify patients suffering from cerebral palsy into a few clearly defined and distinguishable categories suitable for aetiological research. Major categories had to be defined in terms of clinical findings, since living patients were being studied. Clearer definition of categories was obtained by using a number of clinical findings rather than single signs such as an increase or decrease of muscle tone as suggested by Phelps. For example, his category of "Spasticity" includes patients who would be placed in categories of "Hemiplegia", /

"Hemiplegia", "Bilateral Hemiplegia", "Diplegia" and "Ataxic Diplegia" in the present classification. Apart from sharing an increase of muscle tone predominantly in the flexor muscles and increased stretch responses, these conditions are very different from the clinical point of view.

In practice the classification has been found to be satisfactory for the purpose of research into aetiological factors. The importance of birth injury varies greatly in different categories. Some categories appear to contain a high proportion of children with congenital malformations. In others there is a high proportion of cases with familial cerebral palsy.

Though not primarily intended as a means of classifying the extent and severity of the functional handicap which results from cerebral palsy, the classification has been found to indicate these. In particular the arbitrary defined stages of cerebral palsy which are stated give a clear indication of the developmental level of neuromuscular function. For example, the statement that a child is in the "dystonic stage of diplegia" implies that independent sitting, independent voluntary use of the hands and accurate positioning of the limbs are impossible. The extent and severity of the cerebral palsy are also clearly indicated. (Ingram, 1955a).

A complaint often made about modern classifications is that /

that they are so different that series of patients from a number of sources cannot be compared when more than one classification has been used. In practice it has been found easier to attempt approximate reclassification of patients in such series in terms of neurological syndromes than by using one or other of the original classifications employed. When reclassification is attempted it is found that the categories of the present scheme which are most difficult to distinguish in other series are "Bilateral hemiplegia" and "Diplegia", when there is tetraplegic involvement. "Ataxia" and "Ataxic Diplegia" also provide some difficulty. It is usually impossible to give any indication of the stage of the cerebral palsy, or its severity in reclassified cases. Nevertheless, a useful comparison of cases derived from different centres using differing classifications can be made. (Ingram, 1955a). (Table 9).

In practice the classification has been found useful in a number of clinics in this country and in Canada, since it does allow some indication of the level of neuromuscular functioning, and thus of prognosis, to be given. It has been severely criticised for laying stress on the early clinical manifestations of cerebral palsy. (Illingworth,1958) It must be admitted that mildly affected patients may not show such marked changes in findings that "stages" can be defined and a minority of severely affected patients show variations in the course of development of their cerebral palsy. /

palsy. Another criticism is that the distinctions between the defined categories are too fine. However, it is easier to combine small categories after initial classification than to attempt to break down larger categories into smaller sub-categories after initial classification has been made. Research into aetiological categories demands that clinical syndromes shall be clearly and precisely defined. Categories defined in terms of single clinical features such as "Spasticity" or "Rigidity" lack this precision and make for false clinical entities. (Balf and Ingram, 1956).

SECTION 2.

The aims, scope and methods of a survey of Edinburgh children suffering from cerebral palsy carried out in I952-I953.

The Scope and Methods of the Survey.

The Scope of the Survey. The survey was confined to children residing in the City of Edinburgh because, since the size of the childhood population with the city was known within narrow limits, an assessment of prevalence was more likely to be accurate. There were other advantages. A high proportion of patients had been born within the city and there was ready access to birth records, especially from maternity hospitals. Patients were accessible and could be examined repeatedly. Doctors, teachers, relatives, hospitals and clinics having knowledge of the patients could more easily be approached than would otherwise have been the case.

The investigator was also fortunate that in Edinburgh there was considerable interest in cerebral palsy and awareness of the condition amongst doctors, especially in the Maternity and Child Welfare Departments and School Medical Service. Thanks largely to the propaganda of the Scottish Council for the Care of Spastics the tendency for parents of children suffering from cerebral palsy to hide them away was much less marked than in many other places. Thus, patients tended to be referred readily to the survey.

Initially it was intended to confine the investigation to children of school age, born between 1938 and 1948, because they were more accessible for examination than pre-school children or children who had left school. Later it became /

became possible to extend it to include children below school age, though ascertainment in this group proved to be more difficult. A small number of children over the age of 15 were also studied, though not included in the assessment of prevalence.

Eventually the survey included all children resident in the City of Edinburgh at the time of the survey who were born between the years 1938 and 1952 inclusive, whether they had been born in the city or elsewhere.

Methods of the Survey. Various methods of estimating the prevalence of cerebral palsy were considered. Clearly a population of more than 100,000 precluded individual examination by the investigator. Sampling survey would also have been very difficult, for assuming that at most 2 patients might be found in every 1,000 children examined, it would require the examination of 15,000 to produce a series of 30 patients suffering from cerebral palsy (British Council for the Welfare of Spastics, 1948). Even this series would have been too small for studies of clinical findings or aetiological factors to have much value.

It was, therefore, decided to attempt to ascertain, directly and indirectly, all the patients suffering from cerebral palsy who were born between 1938 and 1952, who lived in Edinburgh. This would allow prevalence figures to be worked out for the total child population and by year of birth. Since about 200 patients might be ascertained if the investigation /

investigation were successful, the series should be large enough for clinical findings and aetiological factors to be studied in some detail. It was realised that there was a greater chance of children suffering from cerebral palsy being missed by the survey in such a large scale investigation.

Sources of Ascertainment. Information about possible cases of cerebral palsy was sought in three ways. Firstly, all doctors and organisation likely to see or have under their care patients suffering from cerebral palsy were contacted, and asked to refer them for examination. Amongst those who were contacted were School Medical Officers and Nurses, Doctors and Health Visitors in the Maternity and Child Welfare Department, Medical Officers or Principals of 55 private schools in Edinburgh, Paediatric, Neurological, Psychiatric, Neurosurgical and Orthopaedic Surgeons working in hospitals and clinics within the city. The co-operation of the Scottish Council for the Care of Spastics was obtained and free access to patients known to them was granted. Further patients were referred from clinicians engaged on research projects in related fields. Cases were reported by Dr. H. Provis amongst mentally defective children; Dr. C.M. Drillien from her series of prematurely born patients; the late Dr. J.D. Kerr from children with retrolental fibroplasia; Dr. John Thomson from a study on growth; Dr. J.W. Farquhar from children who had suffered from meningitis, and Dr. Charles Balf from children born after abnormal parturition. It was originally intended

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to write to all general practitioners in the city, but out of the first twenty-five who were contacted, and known to have 28 patients in their practices, only seven replied and gave only the names of two patients both of whom were known from other sources. It was, therefore, not considered worth while to circularise the remaining practitioners, though whenever permission to examine their patients was sought, they were asked if they knew of any other children suffering from cerebral palsy.

The second method of ascertainment was by personal inspection of the case notes, and whenever possible, the children who suffered from disorders which might result in cerebral palsy, or were known to be associated with it. Free access was granted to the records of School Medical Officers, Medical Officers in the Maternity and Child Welfare Department, Maternity, Children's, Orthopaedic, Neurological, Neurosurgical, Psychiatric and other hospitals and clinics.

As many records as possible were inspected of children suffering from mental retardation, epilepsy, overactivity, arthrogryposis, talipes equino varus, visual and hearing defects. Records were inspected of patients who had suffered from conditions which might have caused cerebral palsy, including premature children with a history of birth injury, rhesus incompatibility, meningitis, encephalitis, cerebral thrombophlebitis or abscess, head injury. It was not possible to obtain full records of children from the local Fever Hospital /

Hospital so that those who had suffered from the cerebral complications of infectious disease could be accurately ascertained. More than 2,500 hospital case notes were inspected and approximately 6,000 obtained from School Medical Officers and the Maternity and Child Welfare Department.

The third method of investigation was to arrange for the inspection of selected groups of children amongst whom a relatively high prevalence of cerebral palsy might be expected. Thus, all the schools for mentally and physically handicapped children were visited, and institutions for mentally defective children, and a number of private institutions and schools. After a few months during which experience was gained, it was found most convenient and rewarding to watch children at play in order to select those in whom further examination was felt to be desirable. Teachers were questioned about any children with clumsy writing, unsteadiness, or other suspicious symptoms. A visit to class rooms was made whenever possible as involuntary movements were often more evident there than in the playground. A total of approximately 4,000 children were inspected.

Though apparently rather crude and haphazard, this method was found to give quite accurate ascertainment. In one Special School where it was possible to examine 60 children in detail, and in an institution where over 100 children were examined, no cases of cerebral palsy were detected which had not /

not been suspected on inspection previously.

Thanks to the co-operation of school medical officers it was possible to examine most "Possible Cases" at the time of school medical examinations, frequently arranged especially for the benefit of the investigation. In all, approximately 420 children were examined in the schools. In the examinations of the possible group no notes were taken of those found not to suffer from cerebral palsy, their names were merely erased from the list of possibles. Full notes were taken of all those in whom cerebral palsy was thought to be present or in whom it was suspected even though these children were all seen on subsequent occasions.

Other cases were examined in institutions for the mentally defective, private schools of various types, hospitals and clinics, corporation nurseries, and departments of occupational physiotherapy. Not only did this technique of attempting to arrange the first examination of the child away from parents, save the parents (and the investigator) much time, but it also enabled contact to be maintained with a large number of clinicians and therapists and their interest in the investigation sustained. It was an added advantage that the children were seen in surroundings different from those of their own homes on at least one occasion.

A number of patients, especially those under school age, those classified as ineducable and those having home teaching, were examined only in their homes.

Following /

Following the first examination, arrangements were made with the parents to re-examine and take the histories of those in whom cerebral palsy had been diagnosed. Most frequently these children were visited at home, but a proportion were examined at the time of their routine attendance at various clinics, in the Department of Child Life and Health and sometimes in hospital. Even when, for convenience of the parents the child was examined away from home, an effort was always made to visit the home on at least one occasion. It was possible there to observe the family's reaction to the handicapped child, the behaviour of the child at home, and frequently other relatives than the mother were contacted. On three occasions observation of relatives led directly to the recognition of familial neurological disorder which would not otherwise have been evident.

Methods of Examination.

The methods used in history-taking and examination will be described briefly.

History. The history was taken on all possible occasions from the mother because of her much greater knowledge of the child's early, and especially neonatal history. Whenever possible the mother was questioned on at least two occasions, preferably in different situations. The order of history taking, though of necessity somewhat varied according to circumstances was generally as follows.

A brief family history was taken first. During this information /

information was elicited about the number of people in the family, the patient's place in the family and how he compared in achievement and milestones to his siblings.

The patient's first abnormality was noted and the mother's opinion as to the cause of this. This led naturally to questions about pregnancy and delivery and the neonatal course. At this stage questioning became more detailed and a very full history was taken of the child's milestones, growth, handedness, dexterity, intellectual development and ways in whichhis early development and behaviour differed from the normal. Such detailed questioning at this stage allowed one to slow the tempo of the interview and facilited the mother's thought and the investigator's notetaking. Following the history of the child's early development, the history of the appearance of the first abnormality was taken in more detail and the course of the disorder was followed in detail. This part of the history was usually taken more by encouraging the mother to talk than by close questioning; when her flow of talk had stopped then detailed interrogation was resumed using her story as a basis. In practice this involved a considerable amount of direct questioning in order to elicit points which would not seem of relevance to the mother. A frequent example was, "When he came back from hospital did you find any difficulty in washing him which you had not noted before?" "No, of course not, he was just the same". Direct question. "Did /

"Did you do anything to the right hand before you washed it". "Yes, I had to open it, he always held the fingers closed, though not the other hand". Following such detailed questioning about the origin and development of the child's disorder, questions were asked about his behaviour, illnesses, general educational progress and sociability. Details were then obtained about the history of the family with the emphasis on the mother's obstetrical history and the occurrence of other mental or nervous disorder. Depending on the degree of the mother's co-operation, direct questions might be asked about the occurrence of mental defect and aberration in relatives, or this left until further interviews. Direct questions were asked about consanguinity of the parents. Finally a history of the child's treatment was obtained, with the names of doctors, therapists, clinics, schools, teachers and others who had come into contact with the child and permission obtained to utilise any records obtained.

Following this interview with the mother another appointment was made to see her, either at home, at one of her routine clinic visits, or by special arrangement. After the mother had been seen, as many relatives of the child as possible were seen, especially to obtain an accurate history of the mother's relatives.

In all cases attempts were made to obtain details of the mother's pregnancy with the patient, his birth and neonatal period, from additional sources to the history elicited from the /

the mother or relatives. In 98 cases it was possible to abstract full details from notes of maternity hospitals. Since they relied to a greater extent on memory, details obtained from general practitioners, nurses and nursing homes were relatively scanty and less reliable. Occasionally, however, doctors were able to supply very full details of deliveries which had occurred many years before by reference to their notes.

Notes of infant welfare clinics, hospital clinics, wards, general practitioners, therapists and specialists who had examined the children were requested in all cases. School teachers were contacted whenever possible. The information was helpful in many instances in checking the accuracy of the history obtained from mothers and relatives, and in obtaining a picture of the development of the child's disorder as well as giving an account of the actual causal condition.

The examination. The examination of the child was carried out on more than one occasion in different circumstances in as many instances as possible. The advantages of this were that it was possible to assess the influence of changes in environment on the child's behaviour, and to gain the child's co-operation, if not at the time of the first examination, in almost all cases by the time of the second or third. Especially in dyskinetic patients changes in findings were discovered. Abnormalities missed on initial examinations could be recorded.

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The methods and detailed order of examination varied greatly in different cases and circumstances. The child was weighed and measured for height, difference in limb lengths, trunk symmetry, and head circumference. Any skeletal abnormalities and obvious congenital defects were then noted. The child's demeanour, general behaviour co-operation, apparent intelligence, ability to answer questions and respond to commands were commented on briefly. Note was taken of any speech abnormality that was present and his ability in social activities, reading and writing. A brief general physical examination was made routinely and any abnormalities recorded. A detailed neurological examination followed. It included tests of visual acuity and fields, but not detailed testing of hearing, taste or, in young children palatal and tongue movements. The detail of the examination varied greatly with the age of the child, and in practice sensation could be tested fully only in children with mental ages of 7 years or more.

Greatest attention was paid to examination of the motor system, the child's postural reflexes, tendon jerks and muscle tone being noted in great detail because they could be tested in even the youngest.

Some attempt was made, in all cases, to assess the degree of functional impairment suffered by the child and his ability to sit, stand, walk with and without support, steadiness and manipulative /

manipulative ability were tested. In all stances the child's ability to feed himself, dress himself, handle small objects, manipulate large and small objects, write and co-ordinate his movements in practical situations was noted. From this a far better idea of his capabilities was obtained than by giving standard tests like picking up pins or marbles or completing formboards.

Special examinations. Intelligence quotients were not obtained routinely for the purposes of the survey because most of the children of school age had already been tested and estimates of their intelligence were available. A number of children, especially those under the age of 5 were tested specially by psychologists attached to various schools, departments and hospitals. In a number of cases testing had never been performed and was not requested, either because the child was obviously ineducable or because his abilities were very apparently of a very high order.

Routine electroencephalograms were not performed because of the time necessarily lost to children of school age in visiting the Department of Surgical Neurology, where they were performed. In a large number of children they were available however, and in others, where it was felt they would be of value, they were obtained.

Rhesus blood grouping was not routinely performed, but in all cases where any doubt about rhesus incompatibility was felt they were performed on request by the S.E. Regional Blood /

Blood Transfusion Service.

Radiographs were requested in very few cases. This was partly due to the fact that a special journey would have had to be made by the children to have them made. It is much regretted, in retrospect, however, that more radiographs were not obtained especially from the point of view of estimating bone age and of obtaining permanent records of skull abnormalities.

Fortunately from the point of view of the survey, a number of children had been admitted to hospital and air encephalographs obtained. These and other hospital examinations were readily made available to the investigation.

Difficulties in Ascertainment. Difficulties in ascertainment will be discussed in detail when the measure of accuracy attained in the assessment of the incidence of cerebral palsy is considered. The difficulties were less than had been expected owing to the very good co-operation given by clinicians, clinics, schools and hospitals.

SECTION 3.

A review of the literature on the different forms of cerebral palsy and a description of the findings in 208 patients.

CHAPTER I.

REVIEW OF THE LITERATURE

ON THE AETIOLOGY OF CONGENITAL HEMIPLEGIA

Early Clinico-Pathological Studies.

The first serious attempt to relate the pathological and clinical findings in hemiplegia in childhood appears to have been made by Cazauvielh (1827). In his paper "L'agénesie cérébrale" he quotes 12 case histories of patients in whom cerebral atrophy was diagnosed and confirmed at autopsy in 6 of them. Eleven of the cases had hemiplegia dating from infancy, and were probably of congenital origin, though Cazauvielh emphasises the difficulty of obtaining reliable information about the early months of life. He thought hemiplegia was more common in girls than boys and on the right side than the left. He was aware of the fact that hemiplegia might follow acute infectious illnesses in childhood.

From the pathological point of view he classified his cases into two groups - those with "l'agénesie cérébrale" which were the result of congenital abnormality, possibly the result of failure of development of the cerebral arteries; and those with "l'atrophie consecutif" in which evidence of the destruction of cerebral tissue could be demonstrated.

During the next twenty years controversy was centred on the primary cause of cerebral agenesis; whether it could be the result of foetal encephalitis or meningitis, or whether there was primary failure to develop unrelated to any disease processes (von Lallemand, 1834; Henoch, 1842). Series of cases supporting the first theory were published, and one of these /

these includes the first reference to the presence of subdural and subarachnoid collections of fluid at post mortem which it was felt might be the cause of some of the local forms of cerebral atrophy (Duges, 1826). On the other hand, other authors emphasised the possibility that primary agenesis was responsible, and that the findings were really "formes frustres" of congenital anencephaly (Breschet, 1831).

Cruvehlier (1862) thought that the proposed distinction between primary failures of cerebral development and cerebral atrophy resulting from disease was unhelpful and impossible to establish with certainty on the basis of pathological examination in a high proportion of patients. He distinguished seven categories of cerebral atrophy:-

1. Simple atrophy.

2. Atrophy with granular shrunken convolutions and pigmentation indicating previous haemorrhage.

3. Atrophy with alteration of the cellular content of the convolutions with pigmentation.

4. Atrophy with "cartilaginous induration" of the convolutions after chronic inflammation.

5. Atrophy with loss of substance due to ulceration of the brain following red softening (termed 'porencephaly' by Heschl (1859)).

6. The transformation of a part of the hemisphere into very thin membrane.

7. Chronic meningoencephalitis (pachymeningitis) accompanied by underlying cerebral atrophy.

Cotard (1868) tried to simplify the classification of Cruvehlier and recognised three categories of atrophy:-

1. Chronic meningoencephalitis, in which cerebral atrophy was associated with chronic thickening of the meninges.

2. /

2. Atrophy accompanied by shrinking of the convolutions.

3. Atrophy accompanied by loss of cerebral substance - termed 'porencephaly' by Heschl (1859).

Cotard thought it was impossible to relate the pathological findings to specific aetiological factors in the majority of cases, since atrophy was the end result of a large number of different disease processes.

He agreed with the clinical description of hemiplegia in childhood which had been presented by Cruvehlier, but observed that intelligence was not always impaired and thought that aphasia was relatively uncommon. He also noted asymmetry of the cranium to be found frequently and that the tendon jerks were increased on the affected side, and discussed the skeletal dwarfing of the hemiplegic limbs in more detail. He observed that occasionally muscle wasting was found also and related this to the descending sclerosis of motor tracts in the brain stem and spinal cord which has been previously described by Turner (1856).

Amongst the causes of cerebral atrophy causing hemiplegia or idiocy which he considered to be important in infancy and early childhood, Cotard described traumatic encephalitis, possibly the result of birth injury in some cases, cerebral softening, cerebral haemorrhage, meningeal haemorrhage, cerebral venous thrombosis and "infections at the base of the skull" in infancy. He quoted the finding that of 4941 idiots in whom it was possible to obtain details of the birth, one in 22.5 was born by instruments, and he suggested that this

might
~~may~~ /

~~may~~ might indicate that traumatic encephalitis could cause a variety of nervous and mental disorders in childhood (Mitchell, 1862). Cotard was also aware that some cases of acquired hemiplegia in childhood were the result of acute infections, and specifically mentions measles as a cause.

In the course of his thesis Cotard had described with great accuracy, but without naming them specifically, the changes classified as constituting porencephaly by Heschl (1859) and Kundrat (1882). The latter author made a very detailed and exhaustive study of the pathological findings and clinical correlations of porencephaly. He found that the majority of lesions occurred in the distribution of the middle cerebral artery and that hemiplegia was its commonest single clinical manifestation. On the basis of his histological studies he demonstrated that porencephaly never seemed to occur prior to the fifth month of gestation, but that it could be present in stillbirths prior to the onset of labour and without a history of trauma. He thought many cases were due to thrombosis or embolism of the middle cerebral artery. In others cerebral damage might be due to anaemic necrosis in the tissues supplied by the artery secondary to circulatory imbalance resulting from interference with the placental blood supply during pregnancy or labour. The fact that porencephaly resulted in a great many other neurological and mental conditions besides hemiplegia was pointed out by Audry (1892). He also found it more widely distributed in the brain than Kundrat, recording a smaller proportion in the region of the middle /

middle cerebral artery.

During this period increasing attention was paid by pathologists to the causes of stillbirth and neonatal death. Intracranial venous stasis and thrombosis complicating hypoxia were described by Parrot (1873) and Hutinel (1877). They regarded venous infarction as the commonest cause of cerebral softening, though they realised that the soft consistency of the brain in the newborn also rendered it more liable to traumatic injury. Unfortunately their work was largely ignored by contemporary authors interested in birth injury. They were more impressed by German and American work on the pathological results of traumatic birth injury (Weyhe, 1882; McNutt, 1885; Weber, 1886). McNutt considered that subdural haemorrhages, often observed in stillborn children delivered with trauma, were the commonest cause of cerebral palsy. When unilateral, subdural haemorrhage caused hemiplegia, when bilateral, bilateral paresis. Though it was later pointed out by Collier (1924) that of the three cases she presented to support this theory, one was paralysed but showed no meningeal haemorrhage, and the other two showed meningeal haemorrhage but no paralysis, her work met with general acceptance.

Repeated attempts were made from 1860 onwards to correlate clinical and later pathological findings in congenital hemiplegia. The clinical criteria for the probable diagnosis of porencephaly were described by Brissaud (1896), and for lobar sclerosis by Richardiere (1885). Very detailed studies /

studies were made of the clinical features which might indicate the situation of cerebral lesions in children with congenital hemiplegia (Delhomme, 1882; Dejerine, 1891; Charcot, 1894). The uncertainty of pathological diagnosis on the basis of clinical findings was stressed by Jendrassik and Marie (1885). Strumpell considered that almost all the late pathological findings which had been described in congenital hemiplegia could be the result of prenatal encephalitis (Strumpell, 1889).

Further Aetiological Studies.

Clinical studies of aetiological factors in congenital hemiplegia continued to be made. Gowers (1888) found that of 26 patients 16 were first born, and of the remaining 10 6 had been delivered by the breech. He considered "the hazards of childbirth" to be more important than obstetric interference in causing birth injury.

A history of forceps delivery was found by Osler (1889) in 9 out of 15 cases. He thought intracranial damage had resulted from the instrumental delivery in only 3 out of the nine.

Of 21 children in which the birth histories were known, seven had a history of forceps delivery and four of "very hard labour", in the series studied by Lovett (1888). Four cases of instrumental delivery, two of antepartum trauma to the mother, one of premature birth, three of tedious labour and two of maternal ill health were recorded in 22 cases studied by Sachs and Peterson (1890).

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The majority of American and British papers stressed the danger of trauma rather than asphyxia and appear to have accepted the theories of McNutt, Hadden (1884), Ross (1882) and Oliver (1893). In Germany, however, a rather broader view of the aetiological problems involved was apparent. The possibility of hereditary factors being important in predisposing the infants to cerebral lesions was discussed by Wallenberg (1886). This author also threw considerable doubt on the validity of the assumption that meningeal haemorrhage could be held to account for as many cases of congenital hemiplegia as Gowers and Osler believed. In only six of 19 cases of congenital hemiplegia did he elicit a history of prolonged labour or forceps delivery. Embolism of the middle cerebral artery in prenatal life or infancy was believed to occur not infrequently (Abercrombie, 1887; Osler, 1889). Prenatal encephalitis was thought to explain many cases (Strumpell, 1885).

That the importance of meningeal haemorrhage had been overemphasised was also stressed by Freud and Rie (1891) and Freud (1897). They analysed the pathological findings in cases of congenital hemiplegia and demonstrated that a great variety of pathological findings had been reported. Moreover, though they did not doubt that subdural haemorrhage occurred more frequently as a result of traumatic delivery than of normal delivery, many of the cases with meningeal haemorrhage and hemiplegia showed localised cerebral atrophies and porencephalies which could not have been caused by the haemorrhage. /

haemorrhage. They thought that the association of the conditions was not due to cause and effect but to the action of aetiological factors which were important in and common to both conditions. They were unable to agree with the findings of Wallenberg that hereditary factors, particularly alcoholism in fathers and epilepsy in mothers, could be shown to be important in the aetiology of hemiplegia in childhood. Freud noted that Wallenberg had made no distinction between neurosis and psychosis and that he had included relatives suffering from migraine as indications of neuropathic inheritance.

Freud showed convincingly that congenital hemiplegia could be the result of a great many differing pathological syndromes and that these in turn could be caused by a variety of aetiological factors. He demonstrated that whilst congenital hemiplegia was associated with abnormal pregnancy, traumatic delivery and with neonatal asphyxia in many cases the association was not invariable. It should not be concluded either that because an individual case had been born after abnormal parturition this was necessarily the cause of the congenital hemiplegia. He also emphasised that probable aetiological factors were multiple in many cases and that the way in which they caused specific pathological changes in the infant brain were unknown.

More Recent Studies.

As predicted by Freud, further advances in the understanding of the aetiology of congenital cerebral palsy, including /

including hemiplegia, were forthcoming only when much more interest had been shown in disorders of pregnancy and delivery than previously. This interest resulted largely from the realisation by the turn of the century that infant mortality was disgracefully high and that measures to decrease it were essential (Farr, 1869).

The publication of the results of large series of autopsies on stillbirths had revealed that a high proportion of deaths were due to intracranial haemorrhage (Weyhe, 1889; Kundrat, 1890). A number of workers had reported that between 35 and 45 per cent of stillbirths were due to this cause (Spencer, 1892; Archibald, 1909; Warwick, 1919). Unfortunately in a number of these studies there is no clear distinction between subdural, subarachnoid, intracerebral and intraventricular haemorrhages (Irving, 1930; Munro, 1930). As a result of more detailed work it appeared to be established that subdural haemorrhages resulted most commonly from the effects of traumatic labour and delivery (Beneke, 1910, 1920; Holland, 1920, 1922; Greenwood, 1922). In view of the previous work of McNutt which was still very widely accepted, it was inevitable that the causes of subdural haemorrhage should also almost automatically be regarded as the causes of congenital hemiplegia. Thus one of Little's factors, trauma, was regarded as being of very much more importance as a cause of congenital hemiplegia than were his others including asphyxia (Ehrenfest, 1922, 1923).

A number of authors were not convinced, however, that

trauma /

trauma was the all predominant factor in causing intracranial damage and a number of studies showed that intracranial haemorrhage, especially when subarachnoid or intraventricular, could be due to anoxia acting either as a single or contributory aetiological factor (Capon, 1922; Browne, 1929; Craig, 1938; Schwartz, 1921).

At the same time the pathological picture of anoxic stillbirth and neonatal death was becoming more clearly defined and the importance of anoxia as a cause of death became appreciated (Cruickshank, 1930; Potter and Adair, 1940). Work on the causes of foetal and neonatal death resulting from the processes of birth led to renewed study of the disorders of pregnancy, labour and delivery which might be important in causing congenital hemiplegia. As early as 1913 Batten had suggested that the importance of birth trauma as a cause of congenital hemiplegia was being overstressed and that a proportion of cases were due to the occlusion of cerebral vessels in intrauterine life and to unilateral arrest of cerebral development. The importance of the duration of labour and the occurrence of asphyxia rather than of instrumental delivery in good hands as a cause of congenital hemiplegia was emphasised by Sachs (1926).

A most detailed study of the previous literature and of a large series of personal cases of cerebral palsy was made by Ford (1926). He pointed out the difficulties of determining which, if any, of the large number of possible aetiological factors in his cases were important. He found

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an abnormal birth history in 70 per cent of cases of congenital hemiplegia and considered that birth injury (including anoxia) was the most important cause of the condition. Some cases were also due to intrauterine arrests of development and vascular occlusions. In ten cases of congenital hemiplegia in whom autopsy findings were available he found three had evidence of old subdural clots with atrophy of the underlying ocrtex, four had porencephalic cavities, and three had local cerebral atrophies with lesions of the overlying pia-arachnoid.

Similar conclusions were presented by Stewart (1948). He found that atrophic lobar sclerosis, sometimes involving the whole of the hemisphere, porencephaly and pseudoporencephaly were the commonest pathological findings in his cases of adult mentally defective hemiplegic patients, in whom the condition was of congenital origin. He pointed out the difficulty of attributing the pathological lesions to definite aetiological factors in the majority of his cases. Twenty-six out of fifty had difficult or abnormal labour, and twelve had had abnormal pregnancy. He had doubts about the possibility of cerebral agenesis being a definite pathological entity.

Asher and Schonell (1950) found that of 112 cases of congenital hemiplegia in which the birth history was known, abnormal labour had occurred in 71 and there was evidence of birth injury in 19. Of 41 cases with a history of normal labour, there was evidence of birth injury in 5. They thought birth injury was no more likely to be responsible for asymmetrical cerebral palsy than for symmetrical cerebral palsy. A

proportion /

proportion of cases of spastic cerebral palsy, including hemiplegia, were due to genetic or intrauterine factors.

The aetiological factors involved in various types of cerebral palsy were also reviewed by Yannet (1944) and McGovern and Yannet (1947). The authors had previously confirmed observations that genetic factors were likely to be of importance in the aetiology of cerebral diplegia, and that the significance of birth injury as an aetiological factor had been overstressed. They divided cases of asymmetrical cerebral palsy, the majority of which suffered from hemiplegia, into two groups; the first were those in which a history of birth injury had been elicited; the second were those in which no history of birth injury had been found. The group with a history of birth injury was found to contain a higher incidence of first born children, to have mothers of a more advanced age, to have smaller heads and a lower incidence of prematurity than the group with this history. The latter group was found to show many of the associations also found in cases of congenital malformation, diplegia and mental deficiency (Murphy,1947; Penrose, 1939). Yannet concluded that similar genetic and prenatal factors as operated in these conditions might be important in the aetiology of a proportion of cases of congenital hemiplegia. Birth injury appeared to be more important in congenital hemiplegia (being present in 48%) than in any other form of cerebral palsy. This finding is contrary to that of Asher and Schonell.

The /

The majority of recent British and American studies adopt the classification devised by Phelps which groups cases of hemiplegia and some cases of diplegia together in the category of 'spastic paralysis' (Evans, 1948; Hellebrandt, 1952; Anderson, 1952; Faber, 1947; Denhoff and Holden, 1951). It is difficult to assess to what extent aetiological findings for the group 'spastic paralysis' may be considered to apply to cases of congenital hemiplegia. Certainly no firm conclusions based on these studies can be made.

CHAPTER 2.

REVIEW OF THE LITERATURE

ON THE AETIOLOGY OF ACQUIRED HEMIPLEGIA

Early Writers.

Von Heine (1860) discussing the differential diagnosis of spinal paraplegia in childhood described in some detail the condition of hemiplegic paralysis and gave the first accurate account in modern terms of the onset of acquired hemiplegia, or "acute infantile hemiplegia" as it became known, giving two cases, one following scarlet fever and one vaccination. Being concerned with the differential diagnosis of spinal paralysis, von Heine laid stress especially on the occurrence of convulsions, mental impairment and speech disturbance in hemiplegic patients, for these clinical features suggested brain involvement.

Benedikt (1868, 1874) knew that a large proportion of hemiplegias in childhood occurred as a result of infectious disease. He presented cases following diphtheria, scarlet fever, whooping cough and measles, but was more interested in the uses of electrical stimulation in the diagnosis and treatment of paralysis than in its aetiology.

Richardiere (1885) recorded two cases of acquired hemiplegia complicating typhoid fever, two complicating measles, and one following scarlet fever and vaccination respectively. He described the typical course - fever associated with the sudden onset of recurrent convulsions and the appearance of hemiplegia thereafter, either immediately after the first convulsion /

convulsion or after a delay of a few days. He noted that spasticity appeared in the affected side only after from one to five months, that the fits gradually decreased in frequency over the years and that there was always mental impairment. He attributed all the cases of acquired hemiplegia to lobar sclerosis.

Cerebral atrophy following measles and vaccination was also recorded by Jendrassik and Marie (1885). In the same year cases of acute hemiplegia complicating typhoid, pertussis, measles, smallpox and mumps were reported by Marie (1885). He emphasised the high proportion of patients with relatives who showed evidence of neurological disease, mental deficiency or mental instability, and suggested that some of the patients might be predisposed to cerebral complications of infectious disease by hereditary factors. He also suggested that younger children were more liable to suffer than older children, and that protracted illnesses were more often associated with acute hemiplegia than those of brief duration. He favoured pathological changes in the cerebral arteries, especially in the middle cerebral artery and in branches as the common cause of the condition.

Wallenberg (1886) managed to collect reports of four patients who suffered from acute hemiplegia during the course of typhus, three during whooping cough, thirteen during scarlet fever, nine during measles, two during vaccination, and six attributed to epidemic meningitis (some of the latter on rather dubious grounds) from the literature and from personal observation. /

observation.

Ross (1882) gave "localised haemorrhage or softening" in the hemisphere due to direct cerebral damage, embolism, or thrombosis as the cause of hemiplegia occurring after birth. He emphasised that lobar sclerosis, porencephaly and cerebral atrophy were late appearances of little aetiological significance. Like Ross, Gowers (1888) differentiated between congenital hemiplegia and acquired hemiplegia. He considered that it was commoner in girls and that three-fifths occurred in the first two years of life. In 50 of 80 cases of acquired hemiplegia it was "apparently primary" (in other words, not associated with other apparent infectious disease or other causal factor). He noted the relationship to trauma, diarrhoea, scarlet fever, measles, mumps, whooping cough, bronchitis, pneumonia, and observed that it was more liable to occur in protracted infectious illnesses. He discussed the pathological findings in some detail and emphasised the likelihood of vascular factors being very important on the basis of the clinical and pathological evidence available. He recognised that in some cases the initial lesion was probably an arterial thrombosis, rarely an embolism, but stated that the paresis was much more often spreading venous thrombosis.

Cases in which the actue hemiplegia was attributed to cerebral emboli from endocarditis complicating scarlet fever and diphtheria were reported by Taylor (1880) and Abercrombie (1888), Heubner (1883) and Wiener (1883), but it was pointed out by Osler that "in the great majority of cases the lesions supervene /

Table IO.

The causes of acquired hemiplegia in 94 cases described by some authors before I890.

Author.	Richardière.	Marie.	Wallenberg.	Osler.	Sachs and Peterson.	Total Cases.
Date.	I885	I885	I886	I889	I890	
Cause.						
Typhoid.	2	I	0	0	I	4
Typhus.	0	0	4	0	0	4
Pertussis.	0	I	3	3	4	II
Scarletina.	I	0	I3	7	3	24
Dysentery.	0	0	0	I	0	I
Measles.	2	I	9	4	2	I8
Smallpox.	0	I	0	0	I	2
Vaccinia.	I	0	2	I	I	5
Mumps.	0	I	0	0	0	I
(Epidemic) Meningitis.	0	0	6	0	2	8
Pneumonia.	0	0	0	0	6	6
Trauma.	0	0	0	4	6	I0
Total	6	5	37	20	26	94.

supervene before the onset of these afflictions with which endocarditis is associated". He was able to find only seven cases of cerebral embolism amongst 90 autopsy reports in the literature which described patients who had suffered from acquired hemiplegia in the course of infectious diseases. In his monograph very fine clinical descriptions are given of acute hemiplegia appearing in the course of whooping cough (3), scarlet fever (7), dysentery (1), measles (2) and vaccinia (1). Like most authors except Gowers he found males more commonly affected than females and agreed that the vast majority of cases occurred in the first three years of life (Osler, 1889).

In a series of 83 patients with acquired hemiplegia it was found that four occurred during pertussis, three during scarlet fever, two during measles, two following meningitis, six pneumonia, and one following typhoid, smallpox and vaccinia. (Sachs and Peterson, 1890). They also reported six cases following head trauma, 22 after convulsions of unknown cause, one following tonsillitis, another appearing after an epileptic fit, and one complicating gastroenteritis. In 26 no cause could be stated. The authors emphasised the similarity in the clinical course of patients suffering from acquired hemiplegia whether or no it had occurred as a complication of infectious disease or other febrile condition. They felt that the importance in the aetiology of arterial embolism, thrombosis and cerebral haemorrhage had been underestimated.

The work of Wuillamier (1882), which has been mentioned, was /

was of great importance in furthering the understanding of the sequence of events in acute infantile hemiplegia, and it was he who gave the first detailed account of the gradual change from the flaccid hypotonic paresis immediately after the onset of hemiplegia to the spastic and later contractured stages of the condition.

The clinical studies of all these authors had resulted by the year 1890 in a fairly generally accepted clinical picture of what comprised acute infantile hemiplegia. A child aged under three years, either male or female was affected. There was fairly frequently a history of mental deficiency, epilepsy, mental instability or alcoholism in the family, and there might be a history of abnormal birth, but the child's previous health had usually been good. Either in the course of an acute febrile illness, most commonly scarlet fever, measles, pertussis, or pneumonia, or without any previous prodromata, there was a sudden onset of convulsions. These might be unilateral or generalised, but were usually severe, prolonged and accompanied by loss of consciousness. Following the cessation of convulsions there might be a prolonged period of severe depression of consciousness, but this finding was not constant. Fever might or might not persist for several days or even longer. When consciousness was regained after the convulsions the affected limbs and face (most commonly the right) were found to be paretic and flaccid, and sometimes oedematous. Hemianopia was frequently found and also aphasia if the dominant hemisphere were affected.

It was usually transient.

The paresis was usually most marked immediately after the convulsion and thereafter tended to improve gradually over a period of months with decreasing rapidity for about one year. In a proportion of cases almost full restoration of function occurred, but more commonly there was some persistence of paresis, accompanied within a week or two by spasticity, and within one to three months by the beginning of contracture athetosis, trophic changes and retardation of growth compared to the contralateral limbs. Jacksonian or generalised epileptic attacks continued to occur in over half the patients but tended to decrease in frequency and severity in many cases, most commonly about two years after the acute episode. The degree of mental impairment suffered as a result of the acute episode varied greatly, but might be severe.

It was generally agreed that whether the hemiplegia appeared as a complication of a febrile illness or not appeared to make little difference to its course or its severity. The natural history was the same. But it was noted that whereas in measles, dysentery and pneumonia the hemiplegia most commonly appeared within a few days of the onset of the acute illness, in whooping cough, scarlet fever and diphtheria there was more commonly an interval of at least a week before paresis occurred.

In marked contrast to the more or less general unanimity about the salient points in the natural history of the condition, /

condition, however, was the controversy which raged about the underlying clinical pathology, and this deserves some review.

One of the difficulties which confronted the pathologists attempting to elucidate the nature of the pathological changes underlying acquired hemiplegia was the poverty of pathological material in the acute state. The majority of reported autopsies were on patients who had died from cerebral complications in the course of various infectious diseases, but who had by no means all definitely suffered from acute hemiplegia, and on those whose acute episodes of acquired hemiplegia had occurred many years before.

The latter group was the most studied and the pathological states which were defined have been described (Pages 62-74) The important categories were those of porencephaly, atrophic lobar sclerosis and atrophy with pigmentation. That these findings represented the very late stages of a healing process in damaged cerebrum was not realised for some years, and in the meantime strenuous efforts were made to construct disease entities consisting of characteristic clinical histories and findings on examination to go with each pathological "entity". Thus Richardiere (1885) argued strongly for a disease entity of atrophic cerebral sclerosis. It was suggested that the clinical course of atrophic lobar sclerosis differed from that of the other pathological conditions in that recovery from the initial hemiplegia was more rapid and convulsions were less liable to occur in the first week or two after the acute onset of hemiplegia (Bourges, 1893; Wulliamier, 1882). Brissaud (1896) /

(1886) reviewed this work with favour and attempted to define clinical criteria for the "probable diagnosis" of porencephaly.

At the same time as these writers were attempting to construct disease entities on the basis of pathology, others were attempting to construct them from the clinical findings alone. Vizioli (1880) was impressed by the similarity of the clinical picture in cases of acquired hemiplegia and also by the fact that there was no evidence of pyogenic infection in the brains of the patients he had examined pathologically. He rather tentatively put forward the suggestion that some form of non-septic inflammation might occur in brain which was similar in type to that affecting the cord in poliomyelitis. Unfortunately his clinical descriptions were poor, his pathological findings were questioned, especially in Germany, and his language was Italian. His work was not taken very seriously by either the German or the French authors, and was ignored by the English and the American.

A similar line of thought was being pursued by Strumpell (1885) in Germany, however. He pointed out that cerebral lesions had in fact been observed in poliomyelitis and suggested that an acute non-septic infectious process attacking predominantly the cells, and therefore grey rather than white matter in the brain, would explain many of the otherwise puzzling clinical and pathological features of acquired hemiplegia. He stressed that the diversity of the final pathological lesions was no argument against his theory for their differences might well depend on the severity, locality and /

and timing of the invasion of the aseptic infection. "Primary encephalitis", whether complicating pre-existing infection or not, was merely polio occurring in the brain instead of the cord, though in some cases the cord might also be affected. "The usual form of spinal paralysis, acute poliomyelitis, is known to you. It is no longer in doubt that this is an acute inflammatory disease affecting the grey matter of the spinal cord and the initial stages of this inflammation correspond to the first stage of the disease from which the sequels arise. These sequels depend on the initial defect and are not due to progressive disease. The acute encephalitis of childhood is similar in all particulars, in onset, allowing for the different localisation of the disease process."

Strumpell did demonstrate, even to Freud's satisfaction, that there was a type of acute non-septic encephalitis which could result in hemiplegia, and in a later paper had modified his views sufficiently to acknowledge that occasionally other factors,(cerebral embolism, haemorrhage and thrombosis)could occasionally result in hemiplegia in childhood. (Strumpell, 1891).

His contention that the hemiplegia was the result of "poliomyelitis of the brain", however, was based neither on accurate clinical nor pathological examination. As Freud (1897) remarked, "The result of Strumpell's work was that the medical world was completely taken in and thought he had discovered a new disease." As a result of his work, numbers of cases of "Strumpell's encephalitis" were published in various /

various countries (Williams, 1899; Parkes Weber, 1899). In addition cases were recorded where the two conditions, crebral and spinal, co-existed, though these were rare (Lamy, 1894; Redlich, 1894; Bayer, 1895; Ganghofner,1895). Strumpell (1885) and Moebius (1884) recorded cases where poliomyelitis and hemiplegia had occurred in the same locality or family.

The persistence of Strumpell's ideas may be measured by the fact that three different cases of acquired hemiplegia in childhood in this series have been diagnosed by different clinicians as "Strumpell's encephalitis".

Wallenberg (1886) and Bernhardt (1885) were unable to accept Strumpell's conception of acute polioencephalitis in childhood as the most frequent cause of acquired hemiplegia. The latter dismisses Strumpell in a paragraph, referring to the work of the earlier pathologists, especially Kindrat, in showing how varied the cerebral pathology in cases of hemi-;legia in childhood could be to refute his opinions. Bernhardt (1885) thought venous thrombosis with recanalisation probably occurred frequently.

Freud (1897) discussed the whole subject of cerebral palsy in childhood and correlated pathological and clinical findings to what was known of aetiological factors to the limits possible without the knowledge of the physiology of the nervous system, which were soon to become available. He emphasised the dangers, especially in hemiplegic paralysis, of "making pathological and clinical entities of the types of cerebral /

cerebral palsy". His critical analysis of the previous attempts, especially by French authors, to do this is superbly argued and convincing. He reviewed the whole range of suggested aetiological factors in hemiplegic paralysis in childhood and compared the previous published series of cases point by point. Thus he is able to state that in about one third of cases of acquired hemiplegia in childhood the lesion occurs at the time of an infectious disease, or shortly afterwards.

Bernhardt (1885) reviewed a large number of cases occurring after various infections, from the literature and from his own experience. He pointed out that there is tremendous variety even amongst cases showing similar hemiplegia with similar history arising in the same form of infectious disease and instanced the variability of the pathological findings in whooping cough as examples of assuming too much uniformity in lesions merely on the grounds of clinical likeness.

He thought that it is frequently unwise even to label cases as congenital or acquired, as "one cannot exclude the possibility that all extra-uterinely acting causes, trauma, inflammatory or infectious disease, need the a dded effect of congenital conditioning for them to result in cerebral palsy." He instanced the liability of congenital syphilitics to cerebral vascular lesions during infectious diseases or trauma, and quoted fully cases by Blocq (1894), Gilles de la Tourette (1896) and Erlenmeyer (1891).

Similarly /

Similarly he emphasised that Strümpell's encephalitis involved so many different pathological findings that it could not be regarded as a pathological entity. Since Strümpell's description of clinical findings and especially the onset of the disease, could apply to the majority of cases of acute hemiplegia in childhood, it could not be regarded as a clinical entity either. Freud's destructive analysis of the fallacies, inconsistencies and vagueness of Strümpell's reasoning and the presentation of his case is decisive and complete. This reconciles one to some extent to its ruthlessness and excessive length.

The work of Freud marked a turning point in the history of the literature on acquired hemiplegia. He summarised and assessed the work which had preceded his own, but also looked to the future. In particular, he stressed the need for further knowledge of the physiology of the nervous system before correlation of pathological and neurological findings could be adequate. He also thought that much closer clinical study of patients in the acute stages of their illnesses, which resulted in acquired hemiplegia, would be rewarding, but emphasised that many questions would remain unanswered until more precise diagnostic techniques were developed.

Radiography, advances in neurosurgery and pathological techniques /

techniques, the development of lumbar puncture, air encephalography and immunological and biochemical studies have largely been responsible for the increased understanding of the nature of acute hemiplegia in the past sixty years. It is interesting to speculate about the use to which Freud would have put them had his interest in the subject been maintained.

MORE RECENT WORK.

By the end of the nineteenth century, and largely due to the influence of Freud, it was generally accepted that a large number of different disease processes could cause acute infantile hemiplegia. Aseptic encephalitis (polioencephalitis), cerebral arterial thrombosis, cerebral haemorrhage and embolism, venous thrombosis, cerebral abscess, septic meningitis and cranial trauma were all listed in most works dealing with the condition, though the relative importance attached to each condition by different authors varied widely, as has been indicated.

In the present review the development of modern concepts of the important aetiological categories will be traced from the time of Freud onwards. No pretence is made of a comprehensive review of the literature, but important contributions to the development of current thought will be described.

Virus Infections.

Poliomyelitis. In spite of Freud's arguments against there being such an entity as "Strumpell's encephalitis", clinicians were still keen to discover cases of encephalitis occurring /

occurring in association with poliomyelitis. That small cerebral lesions occurred frequently in patients showing evidence clinically only of spinal involvement had been known from the time of Medin (1898) and confirmed by many observers including Wickman (1913). Moreover the clinical pathological involvement of the brain stem had also been studied and Wickman (1913), Hoffman (1904), Zappert and Spieler (1909) and Peabody (1913) had recorded a considerable number of patients with ataxia as the result of brain stem lesions.

In various epidemics studies were made of residual effects and the conclusions of Wickman (1909), Peabody (1912), Rothman (1931) and Kinnier Wilson (1940) were that though a few of the cases with hemiplegia which were reported might be cerebral forms of poliomyelitis, the condition was extremely rare. It is probable that patients showing lower motor neurone lesions as a secondary result of lateral tract sclerosis in cases of hemiplegia has been mistaken for the occurrence of poliomyelitis in a number of cases (Lamy, 1894; Moebius, 1884; Römer, 1931).

In cases of acute encephalomyelitis complicating measles, for example, the combination of neurological signs indicating the presence of both cord and cerebral lesions might well be misinterpreted as being due to poliomyelitis, especially during an epidemic of the latter. An example of this is included in the present series (Case 24).

Certainly the prevailing opinion now is that though cerebral oedema and focal areas of neuronal damage in the cerebrum /

cerebrum are not uncommon in some epidemics of poliomyelitis, permanent hemiplegia attributable to the virus of poliomyelitis is exceptional.

Other Forms of Virus Encephalitis. In more recent years intensive study has been devoted to other virus encephalitides. Initially this was largely stimulated by the great outbreak of encephalitis lethargica at the time of the First World War first recognised by von Economo (1917) and described more fully by him, fortunately in translation, in 1931. The occasional occurrence of hemiplegia following this condition has been noted, but it is rare (Economo, 1929; Abrahamson, 1935; Neel, 1932). In most of these patients the hemiplegia is associated with mental impairment and other neurological disturbances and is not the predominant residual disability.

Other forms of epidemic virus encephalitis ("diffuse encephalitis" Radermecker) have been responsible for occasional cases of hemiplegia in childhood, including Japanese type B encephalitis (Lewis, 1947; Bredeck, 1938), Australian X disease (Perdrau, 1936). But these conditions do not appear to be important in this country at the present time.

Eastern equine encephalitis, of which there was an outbreak in 1936 and another in 1956 in the Eastern United States, is very frequently a fatal condition (65% in the 1936 epidemic), but the author was privileged in the Autumn of 1956 to see at the Children's Medical Center, Boston, a girl whose predominant residual disability was a left hemiplegia. The diagnosis in this case was confirmed by isolation of the virus from the cerebro-spinal fluid and there was a consistent rise in /

in antibody titre in the blood. This form of encephalitis has not so far been reported in Great Britain.

With the development of virological techniques the identification of increasing numbers of viruses which cause meningoencephalitis, encephalitis and encephalomyelitis has become possible. Some of these, such as the Coxsackie virus, the virus of herpes zoster and sometimes the virus of herpes simplex give clinical indications of their presence, or probable presence, but in the majority of cases identification and confirmation of the nature of the virus disease rests with the virologist. For this reason the number of cases of virus encephalitis attributable to specific viruses is still relatively small and the proportion of each type which suffers from permanent hemiplegia cannot yet be estimated (L'hemitte, 1950; Radermecker, 1956).

Certain types of acute virus encephalitis are known to be much more prone to have permanent sequelae, especially hemiplegia, than are others. For example, the meningo-encephalitis caused by the virus of lymphocytic choriomeningitis (Armstrong and Lillie, 1934) is relatively seldom followed by sequelae attributable to cerebral damage. On the other hand, meningo-encephalitis due to the virus of infective mononuclosis and some of the types associated with preceding virus infections do leave permanent sequels in a number of cases (Kissell and Arnould, 1952; Nilsby, 1953).

Particularly liable to cause marked focal cerebral damage with a high mortality and incidence of persistent neurological abnormalities /

abnormalities are those viruses causing necrotising encephalitis. The virus of herpes simplex is one of these which has been much studied by French authors (Cambier, 1955; van Bogaert and Radermecker, 1955). Occasionally necrotising encephalitis may also complicate herpes zoster (Schmidt, Roseman and Steigmann, 1955).

In other cases, however, though the neurological complications and pathological findings are similar to those found in these conditions, very few positive virus identifications have been made. From the point of view of acute infantile hemiplegia this is unfortunate, for it is in this group of "polioclastic encephalitis"(Greenfield, 1950) or of "necrotising encephalitis"(van Bogaert and Radermecker, 1955) that the clinical course most consistently resembles the clinical course of acute infantile hemiplegia. As has been mentioned, the tendency to focal areas of cell destruction in the brain also makes more likely the occurrence of hemiplegia than in some of the other types of virus encephalitis (Radermecker, 1956).

In the past twenty years another form of encephalitis has been recognised which has a subacute and usually progressive course with remittent fever, hallucinations, periodic regular myoclonus, and occasional attacks of generalised or focal epilepsy are prominent clinical features. Gradual intellectual and emotional disorganisation occurs in association with progressive motor impairment. This condition has been called subacute sclerosing encephalitis and subacute inclusion body /

body encephalitis (Dawson, 1933, 1934; van Bogaert, 1945; Greenfield, 1947). It seems probable, however, that a number of slightly differing clinical pictures have been included in this title (Greenfield, 1947; Brain, 1947).

In the course of this work two cases, one of the van Bogaert type and another resembling a case recorded by Brain (1947), were encountered, but neither has been included in the series as both were resident outside Edinburgh. In the first case the child's general condition mentally and physically is gradually deteriorating, and she has shown for almost two years a mixed picture of hemiplegia and periodic dystonic movement. In the second case with typical clinical and electroencephalographic findings the child recovered but he is left with some mental impairment and a slight left hemiplegia. The numerical importance of cases of this type of encephalitis as a cause of hemiplegia is difficult to assess in retrospect, but seems unlikely to be great.

"Parainfectious Encephalomyelitis and Related Syndromes" (Miller and Stanton, 1956).

Further understanding of the cerebral complications of infectious disease was dependent upon there being available a sufficient number of carefully studied cases for analysis. Gradually there were forthcoming.

The histological findings in the brains of children dying as a result of post-vaccinial encephalomyelitis were reported by Turnbull (1912) and Turnbull and Mackintosh (1926). The pathology of measles encephalitis was studied by

Bergenfeld /

Bergenfeld (1924), Musser and Hauser (1928) and Wohlwill (1928). Three chief pathological changes were reported in the majority of cases - vascular congestion and haemorrhage, especially from the small vessels, perivascular infiltration (rather inconstantly) and perivenous demyelination. The very marked similarity of these findings and the similarities in the clinical course of the neurological disorders in both post-vaccinial and post-measles encephalitis was stressed by Greenfield (1929). He postulated that both conditions might be caused by an unknown virus which spread in epidemic waves but produced no disease unless stimulated to do so by an exanthem. Largely as a result of his work a pathological syndrome of acute disseminated encephalomyelitis, usually occurring as a complication of infections, became generally recognised - a condition termed by Marsden and Hurst (1932) "Perivascular myelinoclasis".

The severe sequels that can attend this form of encephalitis have been increasingly realised. Of 74 cases recently reported, 7 died and at least 20 suffered from mental retardation, personality alteration and paralyses (Appelbaum et al, 1949). The intellectual deficits of children who had suffered from measles encephalitis were studied in great detail by Meyer and Byers (1952). These authors reported that definite impairment of learning ability persisted for a period of months or even permanently in about one-third of the children they studied. They persisted even when physical function and electroencephalograms had returned

to /

to normal. Of the neurological abnormalities after measles encephalitis, hemiplegia and myelitic symptoms are most likely to be permanent and are more liable to occur in the younger child than in the older, according to Miller and Stanton (1956).

A full review of 69 cases of neurological complications occurring in the course of varicella derived from the literature was published by Dagnelie and Dubois (1932). They found that 26% of cases were encephalitic in type and in at least a proportion of cases the pathological reports indicated that "perivascular myelinoclasis" was the underlying process. A fuller review by Underwood (1935) of 120 cases showed that 10% of cases with neurological complications died and another 10% were left with residual paresis or ataxia.

Though aseptic meningitis is a commoner complication of mumps than is encephalomyelitis, it was soon recognised that occasional cases occurred in which the condition of perivascular demyelinating encephalitis could be recognised (Bien, 1913; Larkin, 1919; Wegelin, 1935; Donohue, 1941).

A similar pathological syndrome following "influenza" was recorded by Greenfield (1930) and van Bogaert (1933). Occasional cases following upper respiratory infections due to viruses were reported by de Lange (1939), Bender and Yarnell (1941), and Nilsby (1953). A few scattered reports of perivascular demyelinating encephalomyelitis complicating rubella have been presented by Biggs (1935), Taylor (1937), Davison and Friedfield (1938). Cases have also been recorded complicating smallpox, a variety of other viral diseases, and very /

very rarely scarlet fever (Winkelman, 1942).

The occasional development of perivascular demyelination following the injection of antiserum was recorded by Vogel (1935, 1939), the administration of typhoid, paratyphoid vaccine (Greenfield, 1942; Hughes, 1944; Miller and Stanton, 1954), pertussis inoculation (Van der Horst, 1950) and the injection of diphtheria toxoid (Miller and Stanton, 1954).

In a large proportion of the cases coming to autopsy and showing a pathological picture of perivascular demyelination there was no history of a preceding infection or immunisation, however (Redlich, 1927; Pette, 1927; Martin, 1928; Brain and Hunter, 1929; Radermecker, 1956).

The physiopathological mechanism underlying the occurrence of perivascular demyelinating disease remains in doubt. It was suggested by van Bogaert (1933) that it was in the nature of an allergic antigen antibody reaction. He supported this suggestion with observations of the timing of encephalitic symptoms in relation to the development of various infections and in particular the antibody titre recorded during the illness. He felt that in some families there was a hereditary tendency to reactions of this type. But his suggestion that in measles there is a greater tendency to encephalitis in patients whose skin rashes are mild is not accepted by other authors (Miller and Stanton, 1956). The latter authors examined in detail the available literature on "parainfectious encephalomyelitis and related syndromes" (in spite of Kinnier Wilson's objections to the term 'parainfectious') and also

the /

the neurological complications of immunisation (Miller and Stanton, 1954). They concluded that perivascular demyelinating encephalitis did most probably depend on such a mechanism as was suggested by van Bogaert. They commented on the recurrent nature of the neurological complications in patients receiving repeated immunisations and emphasised that the dangers of proceeding with a course of immunisation if a child should develop even mild neurological symptoms after one or two administrations.

As a result of their comprehensive review of the literature they felt that there was good evidence to suggest that the encephalitis which follows vaccination, immunisation with diphtheria toxoid, typhoid - paratyphoid vaccine, and the administration of antiserum, and which may complicate measles, chickenpox and German measles is usually of the perivascular demyelinating type, though other varieties may occur. The same form of encephalitis is responsible for some of the cases of encephalitis following mumps, occasionally scarlet fever and possibly very rarely pertussis. In the majority of the cases of encephalitis complicating the latter three conditions, however, as in diphtheria, dysentery and gastro-enteritis, other pathological changes not typical of perivascular demyelinating encephalomyelitis are found.

The experimental production in animals of pathological appearances similar to those of perivascular demyelinating encephalitis by injections of antigen and antibody mixtures subcutaneously or intramuscularly, is of great interest

(Morgan /

(Morgan and Kabat, 1947; Kabat and Beyer, 1947).

The finding that a condition of polyradiculitis, similar in type to the Guillain-Barre syndrome in the human, may occur in some animals, and acute disseminated encephalomyelitis in others is highly suggestive that the previous clinical suspicions that both conditions may have an allergic basis may be correct.

Toxic and Haemorrhagic Encephalopathy. "Encephalite Serohaemorrhagique".

A variety of infections have been shown to be complicated by cerebral abnormalities which are pathologically much less specific than the perivascular demyelinating encephalopathy typically found after measles or vaccination. The diseases most studied are whooping cough, diphtheria and scarlet fever which, though relatively rarely complicated by hemiplegia, contributed a considerable proportion of the acquired cases of the condition to most of the early series.

The opinion prevalent in the nineteenth century that the most frequent cause of neurological complications in whooping cough was due to cerebral haemorrhage caused by the rupture of an intracranial blood vessel during coughing was slow to be abandoned, though in fact cerebral haemorrhage is extremely rare (Kinnier Wilson, 1954). On the other hand accounts of areas in which the neurones showed degenerative changes (most frequently in the outer layers of the cortex, the dentate nucleus, and the corpus striatum) of smaller blood vessels showing endothelial swelling and adventitial infiltration /

infiltration and multiple petechial haemorrhages, were presented by Schreiber (1899), Hockinjas (1900), Neurath (1904), Rhein (1905), Hada (1913). Their work was elaborated by other authors, and by 1930 a full complete description of the common pathological findings in pertussis encephalopathy could be obtained from the literature (Husler and Spatz, 1924; Neuberger, 1925; Jochims, 1928). The chief difference of opinion was as to whether the pathological changes in the neurones were due to circulating toxins (Husler and Spatz, 1924), secondary to transitory circulatory abnormalities occurring especially during paroxysms of coughing and convulsions (Heller and Grinker, 1930), or were truly ischaemic in origin being caused by the changes in the blood vessels which could be seen (Winckelman and Eckel, 1927).

Occasional cases in which multiple petechial haemorrhages were accompanied by larger extravasations of blood were also reported (Neurath, 1904; Rhein, 1905; Dolgopol, 1941; Ford, 1952). In these cases of "haemorrhagic encephalopathy" the vascular damage appeared disproportionate to the other pathological findings.

A full review of the literature and descriptions of another 15 cases of pertussis encephalopathy were presented by Dolgopol (1941). She noted the most common pathological findings were, in order of frequency, oedema, eosinophilic neuronal degeneration (homogeneous degeneration of Spielmeyer), multiple petechiae, lymphocytic plugs in the veins and capillaries, small subarachnoid haemorrhages (sometimes with localised /

localised meningeal reaction) and collections of phagocytes and fibroblasts in the vascular adventitia. The non-specific nature of these findings was emphasised by Kinnier Wilson (1954). On the other hand, Miller and Stanton (1954) stress the distinction that can be made between the encephalopathy which complicates whooping cough and that found after scarlet fever or rubella.

Though aseptic meningitis, vascular accidents and septic complications of scarlet fever were recognised to be commoner causes of neurological complications than aseptic encephalopathy, a few descriptions of the latter are available. The changes consist of the same type of neuronal degenerations associated with endothelial swelling and adventitial infiltration in the vessels and petechial haemorrhages, sometimes associated with more extensive extravasations (haemorrhagic encephalitis), as have been described in pertussis encephalopathy. Proliferation of capillary buds was frequent (Toomey, Dembo and McConnell, 1923; Grinker and Stone, 1928; Winckelman and Eckel, 1929). The controversy as to whether these changes were all attributable to the effects of circulating streptococcal toxins or whether the neuronal damage was secondary to the vascular damage was still further complicated by the fact that in many cases neurological abnormalities only became apparent after acute nephritis, septic endocarditis, rheumatism, or middle ear infections had occurred (Ford, 1952).

As in scarlet fever, so in diphtheria, aseptic encephalitis seemed /

seemed likely to be responsible for only a small proportion of the residual neurological abnormalities found after the disease. Degenerative changes in the ganglion cells, usually rather diffuse in distribution, had been observed by Bolton (1903), Globus (1923), and were reviewed by Dolgopol (1950) who also reported a series of eleven cases of her own in which there were no clinical neurological complications. In four of them she found "moderately severe" degenerative changes in the cortical neurones, together with perivascular cuffing in four and microglial nodules in three. She felt that the changes were most easily explained as being due to the effects of circulating diphtheria toxins.

A variety of other infections have been shown to be complicated by similar pathological changes. Typhoid, infantile gastroenteritis, pneumonia, erysipelas, are amongst those in which death has occurred as a result of acute neurological complications, usually high fever, coma or delirium, and frequently convulsions. Pathological examination shows cerebral oedema, vascular congestion, endothelial swelling in the small vessels, frequently some adventitial infiltration and diffuse or focal neuronal degeneration. Petechial haemorrhages are frequent and in a few cases may amount to "haemorrhagic encephalopathy" (Ford, 1952). In passing it should be remarked that it is not known what is the relationship of this form of encephalopathy to that described as "acute necrotising haemorrhagic encephalopathy" (Adams et al, 1949; Russell, 1956). Opinions as to the

significance /

significance of the changes vary widely. Brown and Symmers (1925) thought they were describing a new form of specific encephalitis. Other authors have regarded the neuronal degenerations as the direct result of circulating toxins, and some have emphasised the importance of the vascular damage which is evident in most cases (though not marked by any means in all) (Winckelman and Eckel, 1927; Law, 1930).

In recent years, with the demonstration that very similar changes can occur in states of dehydration or severe metabolic derangements as are found in infantile gastroenteritis, other possibilities are being considered (Alexander and Wu, 1935; Globus, 1932; Crome, 1952). It has been demonstrated that there appears to be a correlation between the circulatory slowing and consequent anoxia in the blood going to the brain, with the level of consciousness and the severity of the pathological changes observed in the brain (Kerpel Fronius et al, 1951). This author considers that the importance of hypoxia occurring in "toxic" states has been underemphasised, especially in the newborn period. Certainly many of the pathological changes observed in the neurones and blood vessels in various toxic states bear a most striking resemblance in type, distribution and course to those described by Courville (1952) as being due to cerebral anoxia. Similar explanations for the cerebral changes complicating pneumonia have been offered by Chorzak (1939).

Venous Thrombosis and Thrombophlebitis.

The complication of thrombosis of the lateral sinus, meningitis /

meningitis and brain abscess have been known to occur in middle ear disease for many years (Abercrombie, 1836; Bouchut, 1884; Passot, 1913; Brunner, 1946). But it was not until the work of Parrot (1873) and Hutinel (1877) that the frequency of the occurrence of venous thrombosis not related to this condition was realised to be a cause of cerebral infarction and haemorrhage in the infant and young child. Unfortunately this work which defined the neuropathological changes found in hypoxia, acute infections, dehydration and marasmus were not sufficiently studied by other workers.

Occasional case reports in the literature did show that other infections than middle ear disease could cause thrombosis of intracranial sinuses and of cerebral veins. Cases following childbirth were presented (Collier, 1891), after broncho-pneumonia (Claude, 1895; Fawcet, 1901; Fisher, 1902), and after scarlet fever and measles (Goodal and Coopland, 1897; Bouchut, 1884).

The most studied group of cases, however, did not consist of children but of mothers showing cerebral complications after childbirth (von Hosslin, 1904; Zangmeister, 1925). As a result of the interest in this subject the pathological and the clinical findings in the condition gradually became correlated to some extent and it was realised that distant infection (in this case in the pelvis) appeared to predispose to the formation of thrombus in the intracranial venous channels.

The importance of venous thrombosis, especially in the young /

young child, as a cause of haemorrhage and cerebral infarction was emphasised by Cobb and Hubbard (1929). These authors presented five cases in which haemorrhage had occurred from congested intracranial veins and capillaries. In three of these there was thrombosis of dural sinuses and pial veins as a result of pneumonia and phlebitis, otitis media and purulent meningitis, and pericarditis respectively.

An attempt to distinguish between dural sinus thrombosis due to infection and dural sinus thrombosis due to dehydration and marasmus and other non-infective factors was made by Byers and Hass (1933). These authors studied 50 cases all under the age of 11½ years, of whom 24 were classified as suffering from "primary thrombosis" (all under the age of thirty months); twenty-six were suffering from a variety of different infections - meningitis (12), mastoiditis (5), cellulitis of the scalp (6), generalised sepsis (3). The commonest organisms in the infective group (excluding meningitis) were staphylococcus aureus and streptococcus haemolyticus.

In the infective group the clinical picture was usually dominated by the preceding infection until signs of meningitis or the onset of stupor, coma or convulsions with preceding headache occurred. In the primary thrombotic group also the condition of the infant often masked the presence of the intracranial lesion. The infants were usually dehydrated, and the longitudinal sinus alone was usually thrombosed without there being clinical evidence of this, but when the middle cerebral veins were involved then unilateral convulsions were ever liable /

liable to occur. As the extent of thrombosis increased further, fits might become more frequent and severe and bilateral, and signs of increased intracranial tension became apparent with stiffness of the neck, bulging fontanelle. The spinal fluid was not very helpful except in meningitic cases. The authors emphasised that the condition of sinus thrombosis might be very silent in its early stages and that there was reason to believe that many patients recovered completely.

The condition of lateral sinus thrombosis in association with middle ear disease was described by Symonds (1931, 1932) as "otitic hydrocephalus". He defined this syndrome as consisting of the symptoms increased intracranial pressure (headache, vomiting and papilloedema) without abscess formation and with spontaneous recovery. In 1937 he described a further five cases in which evidence of raised intracranial pressure but no permanently abnormal neurological signs occurred in association with middle ear disease. He pointed out that the increased intracranial pressure cannot be attributed only to the obstruction of the lateral sinus, and suggested that in the majority of patients thrombus must spread to involve the longitudinal sinus also before thrombosis can occur. He recorded a case of what he believed to be combined lateral and longitudinal sinus thrombosis complicating enlarged glands of the neck. A baby with multiple venous thromboses and longitudinal sinus thrombosis with hydrocephalus due to umbilical sepsis had been recorded by Ellis (1937). Other cases were reported in which "toxic hydrocephalus" complicated /

complicated naso-pharyngeal infections in older children were reported by McAlpine (1937). Three cases, one a child aged six years and the others infants with diarrhoea and vomiting, were reported in the same year in which survival had been long enough for the thrombi in the longitudinal sinus to organise. Both infants showed marked mental impairment after their illness, and all three showed evidence of considerable localised cerebral destruction (Bailey and Hass, 1937). A further paper from the same authors suggested that the frequency of dural sinus thrombosis in infancy had probably been underestimated hitherto and also that thrombosis occurring in states of marked dehydration was very often unrecognised at the time. Many of the so-called traumatic scars found in the brains of children dying from apparently progressive cerebral disease might well be compatible with the occurrence of preceding dural sinus (and especially longitudinal sinus) thrombosis. Cases of cerebral thrombophlebitis were recorded in children with congenital heart disease but without bacterial endocarditis (Lhermitte, Leboullet and Kaplan, 1936). The possible venous origin of cerebral abscesses in congenital heart disease has been reviewed by Ingham (1938), Hanna (1941) and Robbins (1945). The cerebral arteritis and phlebitis found in cases of pneumococcal meningitis was described by Cairns and Russell (1946). They attributed many of the neurological deficits found after pneumococcal meningitis to the cerebral damage caused by this vascular damage.

A total of fifty cases of sinus thrombosis, eight of which /

which were considered to be "marantic" were reviewed by Toomey and Hutt (1949), but only one of the latter occurred in a child, the remainder being elderly adults. Their series of septic thromboses is interesting for the indication it gives of the high proportion of cases with lateral sinus thrombosis in whom the condition has complicated middle ear disease, often with accompanying meningitis, and the frequency with which local infections of the face, nose, nasal sinuses and teeth preceded cavernous sinus thrombosis. Septic longitudinal sinus thrombosis was due to trauma in one case, influenzal meningitis in two, middle ear disease in two, and frontal, ethmoid and sphenoid sinusitis in one.

The whole subject of cerebral thrombophlebitis has recently been reviewed by Garcin and Pestel (1949). Though they recognise the importance of local septic infections of the ears, mastoids, nose and nasal sinuses, face, orbit and scalp in the origin of sinus thrombosis, they stress the importance of general infections in aetiology, and record cases following measles, scarlet fever, bronchopneumonia, gastro-enteritis in the aetiology of the septic variety. Dehydration and wasting diseases in infancy were felt to be the important aetiological factor in the "marantic" type.

Ten cases of acute infantile hemiplegia, in two of whom venous thrombosis appeared to be the cause of the condition, were presented by Mitchell (1952). Others of his cases were probably due to venous thrombosis, and the author concluded that "venous thrombosis, due to minor upper respiratory infection /

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infection may be the cause of many cases of this syndrome (i.e. acute hemiplegia)". Unfortunately the evidence that venous thrombosis had played a part in all his cases is lacking, and the histories, examinations, investigations and course of at least three of them (Cases 1, 2 and 4) are equally compatible with a diagnosis of "toxic" or "perivascular demyelinating" encephalopathy.

Intracranial Abscess.

Whilst the association of intracranial abscess with hemiplegia has long been realised and recognised particularly as a complication of pyogenic ear disease from the early years of last century, interest is now focussed more on the precursors of abscess formation than on its pathology. It has become increasingly apparent that except in patients in whom there is bacterial endocarditis most commonly complicating congenital heart disease in childhood, abscess is most commonly preceded by cerebrothrombophlebitis (Falconer et al, 1943). Even in many cases of congenital heart disease complicated by cerebral abscess the origin appears to be venous rather than arterial (Robbins, 1945; Ingham, 1938; Delille et al, 1938; Lhermitte, 1936). This subject, increasingly important now that congenital heart disease has become more amenable to treatment, has recently been reviewed by van't Hoff (1957).

Thus the most frequent causes of cerebral abscess in childhood are probably now the causes of septic cerebral thrombophlebitis and septic arterial embolism, head injury and osteomyelitis /

osteomyelitis are much less important.

Cerebral Embolism and Arterial Thromboses.

Though at one time cerebral embolism was regarded as being a frequent cause of acquired hemiplegia in childhood, modern opinion is that its importance was probably exaggerated. Thirteen cases of arterial embolism complicating diphtheria and two complicating scarlet fever were found in the literature by Ford and Schaffer (1927). A number of cases of acute hemiplegia followed in some cases by carotid thrombosis palpable in the neck following cardiac catheterisation for congenital heart disease have recently been seen personally. In two of these the children appeared to withstand the procedure well but both suddenly developed predominantly unilateral convulsions between 36 and 48 hours afterwards, and following their fit severe flaccid hemiplegia, which showed very little recovery subsequently, was noted. At the time of the fit in one case and a few hours after in the other carotid pulsation was absent on the contralateral side, and within two or three days the thickened artery was easily palpable. Arteriograph in one case revealed a thrombosis extending to the bifurcation of the common carotid, and no filling of the middle cerebral artery could be obtained from the contralateral side. It is presumed that there was embolism in both these cases and that thrombosis of the artery proximal to the site of embolism occurred.

The importance of arterial thromboses occurring in the course of infectious disease as a cause of hemiplegia was emphasised /

emphasised by Ford and Schaffer (1927), but in the majority of the autopsied cases they record thrombosis as a result of the arterial changes of toxic encephalopathy. They are probably better regarded as cases of toxic encephalopathy rather than of primary arterial thrombosis, though in diphtheria the more specific effect of toxin on the blood vessels may be a commoner cause of thrombosis than is generally realised (Rolleston, 1929).

Occasional cases in which thrombosis occurs without any evidence of any preceding disease do occur. In a recent case a boy aged twenty months gradually lost the ability to speak and the use of first his right arm and then his left leg in the course of 24 hours. On admission to hospital the left carotid artery was not pulsating, and within two days contained palpable thrombus, and a loud bruit could be heard over the right eye. Arteriography revealed a thrombosis extending distal from the bifurcation of the left common carotid. The left anterior but not the middle cerebral artery could be filled from the right carotid. Recovery of function was slight when he was seen about three months later and there was almost complete neglect of the affected side with what was probably a complete hemianopia. He had learnt to walk again, however, and was able to say 'Mummy' and 'Daddy'.

Such cases of "idiopathic" carotid thrombosis must be rare. A more common cause of carotid thrombosis is injury to the carotid artery, either due to the presence of local disease as in a case with septic neck glands recorded by Osler /

Osler (1889) or by trauma in its course. The commonest cause of the latter is a child perforating the tonsillar bed by falling with some sharp object like a pencil in his mouth. Three such cases have been reported recently by Braudo (1956). He points out that though the patients are very often unconscious and may have predominantly unilateral convulsions immediately after the accident the appearance of hemiplegia may be delayed for some hours. In one case seen recently it was possible to confirm the fact that there was absence of pulsation in the carotid for a period of at least four hours before the hemiplegia could be demonstrated. Possibly the brain on the affected side can obtain enough blood to meet its needs from the other side until the means of supply is cut off by the extension of thrombus distally to the Circle of Willis from the site of trauma on the carotid.

Intracranial Trauma.

The mechanisms by which trauma causes acquired hemiplegia in childhood are not as simple as first sight would suggest. Cerebral lacerations resulting from open head injuries do not require a great deal of complicated explanation, though in some cases cerebral abscess rather than the direct traumatic injury is the cause of the hemiplegia (Hood, 1921).

In closed head injury, however, a variety of different processes may cause persistent hemiplegia. These include disruption of cerebral substance as a result of the trauma itself or by bone fragments at the site of injury, by contre-coup, by intracerebral or subarachnoid haemorrhage, and by pressure /

pressure from subdural and occasionally extradural haemorrhages. In addition the persistent hypoxia due to breathing difficulty and sometimes shock and cerebral oedema which so commonly follow head injury with concussion may further impair the vitality of cerebral neurones.

Most attention has been paid in recent years to the syndrome of subdural haemorrhage in childhood. This has been shown to occur most commonly in children under the age of three and of low social class as a result of trauma in almost all cases (Ingraham and Matson, 1941, 1944, 1948). They describe mental retardation as a sequel in approximately 25% of their cases and residual hemiplegia in a smaller proportion, predominantly those in whom there was evidence that the subdural haemorrhages had been present for some months. Though they attribute the persistent hemiplegia in these cases to the effects of severe prolonged and localised pressure on the hemisphere from the overlying haemorrhage it is at least theoretically possible that the hemisphere itself was damaged by the original trauma and occasionally by the removal of very massive adherent membrane at the time or operation. Certainly effective doubt has been cast on the pressure theory in congenital cases of subdural haematoma (Greipentrog, 1959).

Cerebral Hypoxia with Particular Reference to Epilepsy.

That permanent damage to the brain could result from lack of oxygen, or asphyxia, was well known long before the present century (Courville, 1953). A review of the mechanisms of cerebral damage in hypoxia was presented by Barcroft (1920, 1926) /

1926) who was particularly concerned in his later paper with the dangers of high altitude flying.

In recent years the major advances in the understanding of the effects of hypoxia on the central nervous system have been in the definition of the sequence of events and the changes in pathological picture which are found in progressively more severe degrees of the condition (Morrison, 1928; Windle and Becker, 1943; Chorynak, 1939; Courville, 1936; Courville and Marsh, 1933; Courville and Nielsen, 1953; Courville, 1953). In progresively severe degrees of hypoxia the pathological findings were generalised loss of nerve cells, more severe focal loss of nerve cells, laminar necrosis and subtotal cortical necrosis. (These findings will be discussed later with reference to birth hypoxia). Of particular importance from the point of view of hemiplegia was the fact that in many cases the localised distribution of cell necrosis and the prominence of vascular changes were so marked as to suggest, at first sight, that arterial embolism or thrombosis might have played a part. In fact, the final pathological picture following severe hypoxia might be cerebral atrophy, with the opposite hemisphere showing almost normal appearances.

Patients showing evidence of permanent cerebral damage resulting from hypoxia due to carbon monoxide poisoning, strangulation, anaesthesia especially with nitrous oxide, suffocation and other causes were described by Ford (1952), Courville (1936, 1953), Shillito (1936) and Bourne (1955). The most frequently occurring sequelae in children appear to have /

have been visual impairment, epilepsy, educational difficulties and mental impairment, but some cases of cerebral palsy, diplegia, paraplegia and hemiplegia were also recorded (Ford, 1953; Weber, 1931). Thus the question now is not so much whether hypoxia can cause hemiplegia in infancy and childhood but how often it occurs and as a result of what conditions. The current opinion that most of the changes of toxic encephalopathy in the course of severe infections, especially pneumonia, pertussis and gastroenteritis, may be explained on this basis has been mentioned (Kerpel Fronius, 1951; Chorynak, 1939).

A number of recent studies have again described very similar pathological changes to those considered to be typical of cerebral hypoxia by Courville following severe convulsions in infancy and childhood (Zimmerman, 1938; Scholtz, 1951; and Schmidt Crome, 1952; Fowler, 1957). In the majority of these cases there was good clinical evidence of hypoxia at the time of the convulsion. All five of Fowler's cases, for example, had had prolonged convulsions and are noted in his summaries, with one exception, to have been severely cyanosed. It seems highly probable that changes in the nervous system in these patients may be attributed to hypoxia.

If it is true that hypoxia of this degree can occur in severe convulsions it might explain some of the cases in which permanent mental deterioration appears to be the sequel to a particularly severe bout of convulsions in children with recurrent epileptic attacks (Illingworth, 1955). It would also explain the occasional occurrence of hemiplegia after a severe /

severe fit in patients who have had many milder previous convulsions - a sequence of events well known to Wuillamier (1882), Freud (1897), Kinnier Wilson (1953). In these circumstances it would not be too surprising to find that a proportion of patients with acquired hemiplegia were really cases of idiopathic epilepsy who had become severely hypoxic during a fit on account of its severity, on account of inhaling vomitus, or for some other accidental reason.

CHAPTER 3.

REVIEW OF THE LITERATURE ON THE CLINICAL FINDINGS IN HEMIPLEGIA IN CHILDHOOD

By the turn of the century the clinical features of congenital and acquired hemiplegia had been quite fully described. Cazauvielh had noted that paresis affected the limbs more than the trunk, the upper limbs more than the lower limbs and the distal movements of the limbs more severely than the proximal. Fine finger and thumb movements were particularly impaired. Paresis was associated with stiffness of the limbs on passive movements, wasting of muscle and retardation of skeletal growth on the affected side. Incoordinate movements of the affected upper limb, especially the wrist and hand occurred when the patient attempted voluntary movement of these parts. Intelligence was often but not invariably impaired (Cazauvielh, 1827).

Cotard (1868) added the observation that the hemicircumference of the skull was often less over the affected hemisphere than over the non-affected. He recorded that the tendon jerks in the affected limbs were increased and studied contractures and muscle wasting in more detail, relating their occurrence to the presence of sclerosis of the long tracts in the spinal cord. Further observations on skull measurements were presented by Peterson and Fisher (1889). These tended to confirm Cotard's observations though it was apparent that in many cases of hemiplegia the hemicircumferences were approximately equal. The degree of limb dwarfing was studied in

more /

more detail by Féré (1896) and Ferrier (1883). They noted that dwarfing tended to be more severe when the hemiplegia was congenital or acquired in the early months of life than when it was of later onset. Its severity was not consistently in proportion to the severity of impairment of motor function though this was frequently so. The positions in which limbs became fixed by contracture was described in more detail than previously by Brissaud (1880). He felt that severe muscle wasting, associated with what would later have been called lower motor neurone lesions, was not uncommon in hemiplegia in childhood. He attributed the condition to the occurrence of nodular sclerosis in the cord spreading from the degenerated long tracts, which caused pressure damage on the motor cells in the anterior horns. Contractures had also been studied by Little and von Heine, orthopaedic surgoens more concerned with the problem of the surgical treatment of deformities of the limbs than the actual cause of the permanent muscle stiffness (Little, 1853; von Heine, 1860).

The previous rather scattered observations on "Paralysie des nerfs vasomoteurs dans l'hemiplegie" were collected and elaborated by Chevallier and Vulpian (1875). They noted that for a period after the acute onset of hemiplegia the affected limbs tended to be warmer than the unaffected and that there might be oedema in the paretic hand for some days. Gradually, however, the limbs became colder than on the non-paretic side in most cases. Cyanosis and even oedema could result from chilling in severely affected patients.

Involuntary /

Involuntary movements in the affected upper limb, first recorded by Cazauvielh in 1827 were studied in more detail following Hammond's writings on "Athetosis" in 1877. The involuntary movements were quite accurately described by Weir Mitchell (1874) and Gowers (1868). Charcot (1886) attempted to show that they were the result of damage to a single small bundle of fibres in the internal capsule, but this opinion was contradicted by Griedenberg (1886) and Audry (1896).

True athetosis, slow writhing involuntary movements of the fingers when voluntary movements are attempted, was noted by Gowers (1874) to be common in children with hemiplegia. Much less frequently hemichorea might be observed. Gowers emphasised that athetosis was a common complication of hemiplegia and regarded it as such, not as a disease entity as Hammond had suggested.

Hughlings Jackson was interested in children suffering from hemiplegia largely because they frequently suffered from focal epilepsy, which he had described in adult patients. He described the course of convulsive seizures in children with hemiplegia, observing that in many there was a gradual spread of clonic movement from a localised part of a single limb or the face to the whole of that limb, and that the other ipsolateral parts of the body were subsequently involved. In some cases there were characteristic auras. Consciousness might be lost or only slightly impaired. Many patients with a history of having had characteristic "Jacksonian attacks" for /

for a period of months or years would later develop major generalised seizures. Jackson was particularly interested in the focal convulsions because of the evidence they gave as to the site of cerebral lesions (Jackson, 1868).

Wuillamier (1882) extended Jackson's observations on epilepsy in hemiplegia and gave the first fully detailed account of the course of events in acquired hemiplegia. In most cases acquired hemiplegia was first noted after a single severe generalised grand mal convulsion which often occurred during the course of some infectious disease or fever. A smaller proportion of patients had focal or grand mal seizures for a period before they showed hemiplegia and in others the hemiplegia might pre-exist the occurrence of convulsions by months or years. In these latter two groups of patients it was not uncommon to find cerebral lesions which had obviously been present for many years and were frequently clearly of congenital origin. He considered that grand mal or focal seizures were an integral part of the clinical pictures of hemiplegia in childhood. Both Jackson and Wuillamier were impressed by the difficulty encountered in controlling the convulsive tendency in hemiplegic children by the use of drugs. Neurosurgical control of focal fits in hemiplegia was, in fact, attempted as early as 1894 by Angell.

The presence of aphasia in children suffering from hemiplegia was described by Bernhardt and further discussed by Freud, who was at the time writing his classical monograph on aphasia (Bernhardt, 1885; Freud, 1890). They agreed that aphasia was commoner when hemiplegia was on the right than when /

when it was on the left and in acquired than in congenital cases. Difficulties in writing were also noted by Bernhardt and discussed in more detail later by Souques (1894).

Hemianopia was noted to be present in a proportion of patients by Freud (1889) and further discussed by Freud and Rie (1890). Strabismus and field defects were investigated in more detail by Koenig (1894). He found that paralytic strabismus occurred less commonly than simple convergent strabismus in congenital hemiplegia and that actual paralysis of eye movement was commoner in acquired than in congenital hemiplegia.

The onset of puberty in children suffering from hemiplegia was investigated in fascinating detail by Leblais (1894). He recorded cases of asymmetrical breast and testicular development but the clinical significance of much of his work is obscure.

By 1900 large series of patients suffering from hemiplegia had been studied, but these tended largely to confirm the findings of previous workers (Wallenberg, 1886; Osler, 1888; Lovett, 1888; Sachs and Peterson, 1890; Freud and Rie, 1891; Haushalter, 1895). There appeared to be general agreement, however, that both acquired and congenital hemiplegia were rather commoner in the male than the female and on the right than the left side. Mental impairment was generally less than in bilateral forms of cerebral palsy. Aphasia occurred in a proportion of cases and was rather commoner in acquired than in congenital cases.

RECENT WORK. /

RECENT WORK.

Since 1900 great advances have been made in the understanding of the clinical features of hemiplegia in childhood. Increased knowledge of neurophysiology, closer study of the effects of cortical injuries in adults and in experimental animals together with the careful study of large numbers of children with hemiplegia have contributed to this.

The Paresis.

The nature of the loss of voluntary movement sustained as a result of cortical lesions causing hemiplegia has been a subject of controversy since the pioneer work of Hughlings Jackson who wrote in 1870, "The muscles which suffer most are those which can act independently of their fellows of the opposite side ..... parts suffer directly as the actions they engage in are voluntary, and inversely as the actions they engage in are automatic. This is seen in the order of recovery" (from acute hemiplegia). This idea was further expanded in 1874 in his essay on "The anatomical and physiological localisation of movements in the brain" - "Physiologically we say that the patient whose face, tongue, arm and leg are paralysed has lost most voluntary movements of one side of his body, and it is equally important to keep in mind that he has not lost the more automatic movements."

This clear concept of hemiplegia due to brain lesions as causing loss of voluntary movement patterns rather than paralysis of individual muscles was further elaborated by Beevor (1903). He pointed out that muscles which were apparently /

apparently paralysed when some voluntary movements were attempted functioned perfectly well in others and in "automatic movements" such as coughing and sneezing. The nature of the loss of voluntary movements in hemiplegia has been further studied by Walshe (1912, 1922, 1925, 1929, 1943). The movement patterns which are lost are those which tend to be acquired last by the developing child, for example, voluntary abduction of the thumb, extension of the fingers, supination of the forearm are more severely impaired than are voluntary adduction of the thumb, flexion of the fingers and pronation of the forearm (Sherrington, 1906; Ford, 1953). In congenital hemiplegia it is the more complex and later developed voluntary movement patterns which most often fail to appear.

The gradual appearance of hemiplegia in infant monkeys subjected to decorticating lesions showed that it was some time before the classical findings of hemiplegia as seen in the adult developed. Initially there was hypotonia and absence of movement in the limbs on the affected side. Paresis of voluntary movement only became apparent when the child had reached the stage of making wilful voluntary movements. Spasticity took weeks or months to appear (Kennard, 1936, 1938; Tower, 1940). Very similar observations were made on infants suffering from congenital hemiplegia (Byers, 1941). Many hemiplegic babies had been diagnosed as suffering from lesions of the brachial plexus, for the affected upper limbs moved less than the contralateral upper limbs and were flaccid for a period of weeks before true spasticity appeared.

The /

The changes in findings observed when children with acute hemiplegia begin to recover appears to be very similar to that observed in adults (Twitchell, 1951). After initial flaccid paralysis reflex activities of the limbs become apparent and only gradually are these subordinated to a greater or lesser extent to voluntary control. The degree to which voluntary movements are regained appears to be inversely proportional to the intensity of the subcortical reflexes, in particular the grasp and traction responses (Twitchell, 1951).

Reflexes.

The observations of Sherrington (1906) on various decorticate and brain stem animal preparations allowed the activity of the nervous system at different levels to be studied experimentally. The demonstration that higher centres of the nervous system had inhibitory effects on the lower centres and that anatomical levels showed characteristic postural and reflex behaviour was of great importance. It became possible to interpret many of the clinical findings in hemiplegia in terms of the uninhibited action of the remaining undamaged parts of the cortical and subcortical nervous system.

The presence of sucking reflexes, grasp reflexes, withdrawal reflexes, including the extensor plantar response and tonic neck reflexes in hemiplegia were explained on this basis (Walshe, 1923; Walshe and Robertson, 1933).

The occurrence of spasticity and later contracture in the affected limbs was interpreted as being the result of uninhibited cerebellar activity by Jackson (1899). Sherrington thought /

thought that more other subcortical nervous centres also played a part and recognised the importance of vestibular and other brain stem activities. Later work showed that the cerebellum had little influence on the degree of spasticity in hemiplegia (Pollock and Davis, 1927).

The flexed position of the affected upper limb in hemiplegia with an increase in flexor tonus was interpreted as an uninhibited subcortically mediated response to the erect position of the hemiplegic patient by Brain (1927). When the patient was placed in the "quadrupedal position" the affected upper limb assumed a position of extension at the elbow and often the wrist and was capable of supporting the forequarter. It has since been shown that experimentally produced cortical hemiplegia in animals is not invariably accompanied by spasticity and that occasional human patients are found with flaccid cortical hemiplegia (Aring, 1940; Denny-Brown and Botterell, 1948). Nevertheless the principle remains unchallenged that cortical damage removed inhibition from related centres of the nervous system and allowed their activity to be manifest.

Normal infants show many of the reflexes which appear in the adult only when cortical inhibition has been diminished as a result of brain damage. In congenital hemiplegia the grasp reflex, the tonic neck reflex, and less constantly the sucking reflex persist on the affected side instead of being lost as on the unaffected side. The plantar response remains extensor (Byers, 1941; Thomas and Sainte-Anne-Dargassies, 1952; Koupernik, 1954).

Involuntary Associated Movements.

The writhing movements of athetosis found in a relatively high proportion of children suffering from hemiplegia have been interpreted in a number of different ways [redacted]. The commonest explanations are that there is lack of inhibition of, and damage in, the subcortical nervous centres, especially the basal ganglia, concerned with postural control of the limbs during voluntary activity (Thomas, 1932). This hardly explains why athetosis should be abolished by extirpation of the contralateral motor cortex, however (Horsley, 1909).

As a result of detailed study of the grasp reflex in experimental animals and human patients another explanation may be offered. It has been observed that there are a number of components in the grasp reflex and that the affected hand behaves differently when different parts of the contralateral cortex are extirpated. When the frontal lobe is removed the grasp reflex is easily elicited by distally moving pressure in the palm of the hand. When the temporal lobe is removed the tendency is to groping in the affected hand, for the hand to follow moving objects in an attempt to hold them. In parietal lobe lesions the hand often moves away from stimuli and shows an "avoidance response" (Denny-Brown and Seyffarth, 1948; Denny-Brown, 1956; Twitchell, 1958). It has been suggested, though it remains unproved, that the involuntary movements of athetosis may be interpreted as fragments of these various forms of involuntary hand activity occurring out of phase and incoordinately during attempted voluntary movement /

movement of the affected limb (Twitchell, 1958). The involuntary chorea-like movements which occur much less frequently in hemiplegia in childhood cannot be explained on this basis, however, and it seems likely that there is damage to the underlying basal ganglia, as well as to the cortex in these patients.

Involuntary changes in the position of the affected limbs occurring with voluntary activity on the healthy side, with emotion and changes of posture, were studied by Riddoch and Buzzard (1921). The types of involuntary movements observed have been fully classified and described by Monrad Krohn (1948). Generalised associated movements ("syncisesie globale pour spastique") are movements elicited readily by all kinds of exertion and tend to produce the typical attitude of predilection, which is of generalised flexion in the hemiplegic upper limb, when the patient is in the erect position, as described by Brain (1927). Symmetrical associated movements ("syncinesies d'imitation") are imitative movements of the non-paretic side carried out by the affected limbs. Co-ordinated associated movements consist of the involuntary movement of synergistic muscle groups, such as the involuntary dorsiflexion of the foot when the hip on the affected side is flexed, or flexion of the affected hip when the patient is asked to sit up from the supine position without the help of his hands. A number of these movements have been found to be extremely useful in physiotherapy, for they allow joint movements to be produced which the hemiplegic child cannot make voluntarily (Thomson, 1956).

Sensory /

Sensory Abnormalities in the Limbs in Children Suffering From Hemiplegia.

Neglect of the hemiplegic arm in children was noted by a number of the early authors, but apart from a single patient with associated hemianalgesia described by Charcot, impairment of sensation was not observed (Ford, 1952). Detailed testing of sensory functions in 106 patients suffering from congenital or postnatally acquired hemiplegia was carried out by Tizard, Paine and Crothers (1954). Touch, pain, temperature, position size, passive motion, vibration, location sense, sharp dull discrimination, two point discrimination, stereognosis and texture recognition were examined. It was found that sensation, especially kinaesthesia, was impaired in approximately 50% of patients, rather more commonly in those with acquired than those with congenital hemiplegia. Hemianopia was discovered in under 25%, and when present it was invariably associated with sensory defects in the homolateral limbs. The presence or absence of sensory defects did not appear to correlate with the severity of motor dysfunction, mental impairment or the presence or absence of epilepsy. It tended to correlate closely with bony undergrowth of the affected arm. In some patients the sensory defect appeared to be making a major contribution to the uselessness of the affected upper limb. Subsequent authors have confirmed that sensory abnormalities in the upper limbs of hemiplegic children occur frequently (Woods, 1957).

In addition to actual impairment of sensory appreciation in the affected upper limb there is frequently neglect of the hemiplegic /

hemiplegic side, apparently associated with lack of awareness of them and a disturbance of body image. The neglect which is characteristically found in parietal lobe lesions in experimental animals has been much studied in adults with parietal lobe lesions by Critchley (1953) and Denny-Brown (1956). There are great difficulties in assessing impairment of body image in children, but it is evident from recent work that this occurs frequently in children suffering from congenital or acquired hemiplegia, and that it may be a major disability (Russell, 1959). Not only is the child's concept of his own body disturbed but his perception of the environment, his orientation in the environment and his ability to deal with shapes and their relationship one to the other are much impaired (Albitreccia, 1958).

Apraxic phenomena are frequently encountered in children with hemiplegia. Katapraxia (termed the Penelope syndrome by Pineas, 1931) has been observed less frequently. Unfortunately it is much more difficult to analyse the apraxic component in children that it is in adults, especially when the hemiplegia has been congenitally acquired or when patients are mentally retarded. Much more detailed study is required of apraxia in children with hemiplegia than has so far been possible.

Epilepsy.

Surveys of large numbers of children suffering from hemiplegia have resulted in an improved assessment of the frequency with which epilepsy occurs. In Bristol it was found that 43% of 97 hemiplegic patients suffered from fits. These /

These were more frequent in children with acquired hemiplegia than amongst those suffering from congenital hemiplegia (Woods, 1957). Thirty-five per cent of hemiplegic children were found by Illingworth (1958) to be epileptic. Perlstein and Hood (1955) found that 43% of patients had epileptic seizures. In Edinburgh 21% of children with congenital hemiplegia and 46% of children with acquired hemiplegia had convulsions. (Nine of the 24 patients with epilepsy had Jacksonian convulsions, one petit mal only, and the others grand mal.) (Ingram, 1955). Differences in the prevalence of epilepsy indicated by these figures are probably mainly the result of differences in sampling. Whilst the prevalence of epilepsy in hemiplegic patients has been closely studied, less attention appears to have been paid in most studies to the frequency of fits, their duration and clinical type. Yet the finding that hemiplegic children who suffer from epilepsy are significantly less intelligent that are those who do not suggests that further study would be well worth while (Perlstein and Hood, 1954). At least four types of seizure have been described; the generalised grand mal convulsion, the focal or Jacksonian seizure, and less commonly myoclonic jerks and true petit mal. The prognosis for intelligence appears to be worst in patients with myoclonic jerks (Bower, 1959). Patients with only infrequent focal or Jacksonian attacks, especially if they are not accompanied by loss of consciousness, appear to have a better prognosis than those with grand mal and accompanied by loss of consciousness. It has to be remembered, however, that a proportion of patients with only focal fits for /

for months or years may later have generalised grand mal convulsions (Ford, 1926).

In hemiplegic children with very frequent convulsions in which a "trigger point" can be incriminated clinically or electroencephalographically removal of the affected part of the brain (lobectomy or topectomy) or of the whole hemisphere may be performed. The success of this operation in severely affected patients has encouraged study of the fits and encephalographic findings in hemiplegic children. A wide variety of patterns have been recorded varying from diffuse generalised high voltage dysrrhythmia to focal spike, commonly in the temporal or temporo-parietal region superimposed on an otherwise normal looking record. The voltage may be increased and the waves irregular over the damaged hemisphere or they may be regular but reduced in amplitude (Perlstein, Gibbs and Gibbs, 1953). In the small minority of patients with myoclonic jerks either an electroencephalographic picture of hypsarrhythmia or of irregularly occurring waves and spikes may be found (Gibbs,1952).

Behaviour.

Overactive behaviour has long been noted to occur in a proportion of patients suffering from congenital or more commonly acquired hemiplegia, the majority of whom suffer from focal fits or generalised grand mal convulsions (Peterman, 1953). The overactivity has marked similarities to that which may be observed in animals after experimentally induced damage to the hippocampus or inferior portion of the temporal lobe /

lobe (Klüver and Bucy, 1939). Behaviour is typified by an increase in distractability, poor attention span, an apparently irresistible urge to touch and often to chew every object which is in reach, a diminished capacity for spontaneous affectionate behaviour, aggressive outbursts, an absence of fear and a failure to respond to reprimand or chastisement (Balf, 1952).

Behaviour of this type has been described in 70 epileptic children by Ounsted (1955). It may be observed in patients without seizures, however, and was described in 25 cases, 12 of whom had no other evidence of epilepsy, by Ingram (1956). Of these 25 cases 17 were suffering from congenital or acquired hemiplegia. Ounsted's observation that phenobarbitone almost invariably exacerbated the overactivity whilst primidone tended to diminish it markedly in a proportion of patients was confirmed.

In a high proportion of patients with this form of overactive behaviour ("the hyperkinetic syndrome") electroencephalographic foci may be recognised by electrocorticography if not through scalp electrodes. Temporal lobectomy and hemispherectomy have benefitted the behaviour and epilepsy in such patients to a quite remarkable degree. Hemispherectomy has been followed by surprisingly little impairment of speech or motor or intellectual functions (Penfield and Paine, 1953; Penfield and Steelman, 1947; Krynauw, 1950; McKissock, 1953; Meyers, 1955).

Much greater understanding of the psychological stresses faced by children suffering from hemiplegia has resulted from a /

a number of recent studies (Gibbs, 1958).

Aphasia and Impairment of Speech.

The large survey of patients suffering from cerebral palsy recently carried out in Boston showed that there was no significant difference in the rate of speech development in children with right and left hemiplegia (Paine, 1958). Retardation of speech development and speech defects were more frequent in children with severe mental impairment than in those in which it was slight.

The actual prevalence of speech defects in the many recent surveys is difficult to assess, for such different criteria are used. Though severe dysarthria appears to be rare in hemiplegia, minor articulatory abnormalities are common, especially in younger children. Ten point five per cent of hemiplegic children suffered from speech defects in the survey made by Skatvedt (1958). Asher and Schonell (1950) considered speech defects to be rare in hemiplegia. It seems likely that simple retardation of speech development (to be expected in mentally retarded children) is being confused with dysarthria by a number of authors.

Aphasia is commoner in acquired than in congenital hemiplegia and becomes commoner the later in post-natal life the hemiplegia is sustained (Gros and Vlahovitch, 1954). That profound dysphasia does not follow hemispherectomy in children with congenital hemiplegia has been attributed to the failure of the damaged hemisphere to paly a part in controlling speech. But recent studies on the correlation of handedness and the laterality /

laterality of speech function indicate that the problem is a complex one (Meyers, 1958; Subirana, 1958; Zangwill, 1954; Zangwill, 1959; Goodglass and Quadfasel, 1954).

In many hemiplegic children with aphasia there is retardation of speech development rather than striking disorder of language usage. The retardation of speech development is commonly associated with articulatory errors which are of characteristic type, and later with specific dyslexia and dysgraphia. These are much more commonly recognised as disabilities in the hemiplegic child than the inconspicuous dysphasic difficulties which are also commonly found to be present if they are looked for (Ingram, 1959a).

Intellectual Impairment and Educational Difficulties.

Recent studies of large series of children suffering from congenital and acquired hemiplegia have shown that the amount of intellectual impairment varies greatly in the condition. More children with severe hemiplegia will have severe mental impairment than will those with mild hemiplegia, but the severity of the mental and physical disabilities are by no means constantly parallelled. No significant difference was found in the proportions of defective children and children with average intelligence amongst right and left hemiplegic patients (Perlstein and Hood, 1955; Asher and Schonell, 1950). Woods (1957) found that there were more right than left cases of hemiplegia with intelligence quotients under 70, but there may well have been some unwitting selection of cases in her survey. Perlstein and Hood (1955) found no significant difference in the intelligence quotients of children with congenital /

congenital and those with acquired hemiplegia. Overall, patients suffering from hemiplegia had mean intelligence quotients of 20 less than children in the normal population.

In addition to impairment of intelligence there are many other disabilities which prevent the child with hemiplegia making normal educational progress. These have been listed by Schonell as follows:- motor involvement, sensory impairment (visual, auditory, kinaesthetic), speech impairment, faulty visual perception, faulty auditory perception, faulty visuo-spatial perception, laterality complications, perseveration, fluctuating performance, lack of drive, distractability, deprivation of experience, overdependence, frequent absence from school. In many patients suffering from quite mild hemiplegia aphasia, faulty visuo-spatial perception, laterality complications and faulty appreciation of their own body image are major causes of educational retardation. There is persistent difficulty in recognising letter and word shapes, in perceiving the significance of their relationships in space to each other and in interpreting them in phonic terms. These difficulties are almost identical to those encountered in children with "developmental aphasia" (Ingram, 1956; Drew, 1956). The detailed study of children suffering from the after-effects of measles encephalopathy, some of whom were hemiplegic, illustrates many of their intellectual and educational difficulties very well (Meyer and Byers, 1951).

CHAPTER 4a

Aetiological studies of 75 children suffering from hemiplegia.

Introduction.

For the purposes of description, patients were placed in one of three categories, according to the time at which their paresis, or the first signs of neurological abnormality, became apparent. Patients classified as suffering from congenital hemiplegia are those who showed neurological abnormality at the time of, or shortly after, birth. In the majority of the thirty patients so classified evidence of disorders of pregnancy, delivery or neonatal course was present. Patients classified as suffering from acquired hemiplegia were those in whom evidence of an apparently normal neurological state for a period following birth was obtained, and in whom a history of the later onset of hemiplegia could be elicited. The majority of the 33 patients considered to show acquired hemiplegia suffered from acute diseases and their complications, which resulted in the appearance of the paresis. Twelve patients were classified as suffering from hemiplegia of unknown aetiology. In some of these patients full histories were obtained, but it was impossible to determine if, as babies, they had been normal for a period after birth or not. In others the classification was due to inadequate histories being available, either because parents were ignorant, mentally defective, or because they would not co-operate in the survey. Most were of low social class.

Family History.

Parents. No parents of children with hemiplegia themselves suffered from cerebral palsy.

In 7 cases one of the parents suffered or had suffered from recurrent epileptic fits after the age of 5 years. In three cases the father was affected and in four the mother (Table 11). In three cases a history was elicited of other relatives having epileptic attacks as well as the parents. Three of the cases with epileptic parents were of congenital and four of acquired hemiplegia. No parents were recorded as being epileptic amongst the cases of hemiplegia of unknown origin. The prevalence of known epilepsy amongst the 150 mothers and fathers was about 5%. The cause of the epilepsy was unknown in most cases.

In four cases of congenital, three of acquired hemiplegia, and in three of unknown origin, one or both parents were classified as feeble minded. All were in Social Classes IV or V. The prevalence of feeble mindedness amongst parents was approximately 9%, and ten, or 12.5%, of patients had at least one feeble minded parent. Six of these feeble minded parents had given birth to children (siblings of the patients) who were also feeble minded, as indicated in Table 12 .

In case 162 the mother had acquired a hemiplegia following previous parturition. She had developed a "white" leg a few days after delivery and the hemiplegia was attributed to embolism. Of her six children, three were healthy, the patient suffered from congenital hemiplegia and two attended schools /

TABLE 11

Epilepsy and Disorders of the Nervous System Amongst the Parents of Hemiplegic Patients

Case No.	Social Class	Consanguinity	Mother	Father	Siblings	Other Relatives	Stillbirths and Infant Deaths
Congenital Cases.							
27	V	-	Feeble minded	Epileptic	1 sister mentally defective	M.G.M. defective	-
100	IV	-	Healthy	Epileptic	2 healthy	2 paternal uncles epileptic	-
162	V	-	Hemiplegic	Healthy	3 healthy 2 mentally retarded	-	-
192	V	+	Epileptic	Healthy	1 healthy	-	-
Acquired Cases.							
19	III	-	Epileptic	Healthy	1-2 healthy	Maternal aunt epileptic	1 stillbirth 1 neonatal death
48	I	-	Epileptic	Healthy	1 healthy	-	-
52	III	-	Epileptic	Healthy	-	-	1 stillbirth
132	V	-	Tuberculous psychopath	Epileptic	2 healthy	Paternal uncle and paternal grandfather epileptic	-

TABLE 12

Patients with Mentally Subnormal Parents

Case No.	Mother	Father	Siblings	Other Relatives	Stillbirths and Infant Deaths	Social Class
Congenital Cases.						
27	Feeble minded	Feeble minded	1 healthy 1 brother mentally backward	M.G.M. mentally backward	1 stillbirth	V
109	Feeble minded	Feeble minded	1 mentally defective	-	1 stillbirth	IV
111	Feeble minded	Healthy	-	-	-	IV
170	Feeble minded	Healthy	7 healthy	-	-	IV
Acquired Cases.						
89	Feeble minded	Feeble minded	1 mentally retarded 2 psychopathic	-	-	V
184	Feeble minded	Healthy	2 healthy	-	-	V
188	Feeble minded Alcoholic	Healthy	At least 1 out of 7 sibs. feeble minded	-	1 stillbirth	V
Cases of Unknown Aetiology.						
58	Feeble minded	Healthy	3 healthy	-	-	IV
168	Feeble minded	Feeble minded	-	-	-	V
199	Feeble minded	Healthy	1 healthy 1 diplegic	-	-	IV

TABLE 13

Psychiatric Disorders Amongst Parents of Hemiplegic Patients

Case No.	Social Class	Mother	Father	Siblings	Consanguinity	Other Relatives
Congenital Cases.						
39	III	Healthy	Alcoholic	1 diplegic 1 died:- congenital heart	-	-
74	V	Probably psychotic	Healthy	-	-	-
79	III	Anxiety neurosis	Healthy	-	-	-
97	III	Anxiety neurosis	Healthy	3 healthy	-	-
109	IV	Low intelligence Anxiety neurosis	Low intelligence	1 mentally defective (1 stillbirth)	-	Mother's brother and nephew mentally retarded
123	III	Healthy	Psychopathic	-	-	-
167	II	Healthy	Alcoholic	-	-	-
Acquired Cases.						
50	IV	Anxiety neurosis	Alcoholic	4 healthy	-	-
89	V	Low intelligence	Low intelligence and probably psychotic	1 mentally defective 2 psychopathic 1 healthy	-	-
132	V	Psychopathic	Epileptic	2 healthy	-	-
188	V	Low intelligence Alcoholic	Low intelligence	3 poor intelligence 3 healthy	-	Not known
Cases of Unknown Aetiology.						

schools for the mentally handicapped.

Psychiatric disturbances and alcoholism occurred relatively frequently. Five mothers were considered to show definite anxiety neurosis, and three were psychotic, psychopathic and alcoholic respectively. Four fathers were considered to be alcoholic, one probably psychotic and one psychopathic. Seven of the total of 12 cases in which the parents suffered from these disorders were in Social Classes IV or V. (Table 13.)

Siblings. Two patients with hemiplegia, one congenital (Case 39) and one of unknown origin (Case 199) had siblings suffering from congenital diplegia. In Case 199 the patient's diplegic sister was included in the survey. Another sibling was mentally retarded. (Table 14.)

Epilepsy was present in the siblings of two patients, one with hemiplegia of unknown origin (Case 134) and one with acquired hemiplegia (Case 93).

Feeble mindedness was present in 11 siblings of 9 patients. Six of these patients were congenital hemiplegics, one suffered from hemiplegia of unknown origin and two from acquired hemiplegia (Table 14.). In six of the 9 cases one or both parents were mentally retarded also. The numbers of siblings considered to be feeble minded (or worse) is probably an underestimate, for many siblings were under school age and the criteria employed for diagnosis did not apply.

In Case 50 an older sibling, whom it was found impossible to run to ground, was said to suffer from "congenital chorea". In Case 169, also a case of acquired hemiplegia, 3 siblings and /

TABLE 14

Abnormal Siblings of Patients Suffering from Hemiplegia

Case No.	Social Class	Father	Mother	Siblings		Stillbirths and Neonatal Deaths
				Normal	Abnormal	
Congenital Cases.						
5	IV	Healthy	Healthy	1	1 mentally retarded	1 stillbirth
29	V	Mother feeble minded Father epileptic	Healthy	0	1 mentally defective	-
39	III	Father alcoholic	Healthy	1	1 diplegic	1 neonatal death Congenital heart
109	IV	Feeble minded	Feeble minded	3	1 mentally defective	1 stillbirth
162	V	Healthy	Hemiplegic	3	2 mentally retarded	-
170	IV	Healthy	Obese and not very intelligent	5	2 mentally retarded	-
Acquired Cases.						
50	IV	Alcoholic	Healthy	3	1 "congenital chorea"	-
89	V	Feeble minded	Feeble minded	1	(2 psychopathic) 1 feeble minded	-
93	III	Healthy	Healthy	2	1 epileptic	-
169	IV	Healthy	Healthy	1	3 congenital talipes	1 neonatal death
188	V	Feeble minded	Feeble minded	6	1 feeble minded	1 stillbirth
195	IV	Healthy	?psychotic	6	1 epileptic	-
Cases of Unknown Origin.						
119	IV	Healthy	Healthy	3	1 died hydrocephalus	-
134	III	Healthy	Healthy	10	1 epileptic	1 neonatal death
199	V	Healthy	Feeble minded	1	1 diplegic 1 feeble minded	-

and the patient were born with bilateral talipes equino varus deformities. In Case 119 (hemiplegia of unknown origin) a sibling died of internal hydrocephalus at the age of a few months.

Neonatal deaths were ascribed to prematurity in Cases 84, 134 and 19 (a twin), to whooping cough in Case 19 (a twin), and to spina bifida in Case 169. Infant deaths were attributed to "otitis media and pancreatitis" in Case 165, endocardial fibroelastosis in Case 39, and "convulsions" in Case 134.

Taking all 153 live-born siblings of hemiplegic patients, 9 had died, at least 3 of them as a result of congenital anomalies, and 19 who survived were considered to show physical or mental abnormalities of congenital origin.

Other Relatives. In two cases of congenital hemiplegia relatives certainly had cerebral palsy (Cases 29 and 201). In the first the maternal grandmother was diplegic, and in Case 201 the mother's sister's child was diplegic. (Table 15)

A history of mental defect amongst relatives was elicited in Case 29 (in addition to a history of cerebral palsy) and in Case 109 in which the mother's brother was feeble minded and his son mentally defective.

A history of epilepsy amongst relatives was elicited in one case of congenital hemiplegia (Case 100) and in two cases of acquired hemiplegia (Cases 19 and 132). In Case 100 two paternal uncles as well as the patient's father were epileptic. In Case 19 a maternal aunt and in Case 132 the paternal grandfather and a brother of the father were epileptic as well as the father himself.

TABLE 15.

History of Abnormal Relatives of Patients Suffering from Hemiplegia

Case No.	Abnormality in Relatives	Health of Parents	Health of Siblings	Stillbirths and Neonatal Deaths
Congenital Cases.				
29	M.G.M. Probably had cerebral palsy Paternal cousin mentally defective	Father epileptic Mother healthy	1 healthy	
100	2 paternal uncles epileptic	Father epileptic	2 healthy	-
109	Maternal uncle and his sons mentally retarded	Both poor intelligence	1 mentally backward 3 healthy	1 stillborn at 7/12
201	Cousin (mother's sister's child) diplegic	Mother healthy	-	-
Acquired Cases.				
19	Maternal uncle died of convulsions	Mother healthy	11 healthy	Twins (1 stillborn; 1 died neonatal period) 1 death at 3 weeks
132	Paternal uncle epileptic	Father epileptic	2 healthy	-

The prevalence of cerebral palsy, epilepsy and mental retardation amongst parents and siblings.

In Tables 16,17and 18 the patients with parents or siblings giving a history of mental defect or epilepsy are tabulated. In addition, the cases in which other relatives suffered from cerebral palsy are recorded.

It will be observed that 4 patients had a family history of cerebral palsy, the diagnosis being of diplegia in all cases. Since the prevalence of diplegia in the child population is about one per thousand, the occurrence of two cases of diplegia amongst the 144 surviving siblings of hemiplegic patients is remarkable. It suggests that there may be common aetiological factors in the conditions. This possibility will be discussed in more detail later.

Epilepsy was present in the parents or siblings of 9 patients, one of whom (Case 29) also had a family history of cerebral palsy. Convulsions occurred in one parent in 7 cases, and in siblings in two. The prevalence of epilepsy amongst the siblings is probably not more than would be expected in the child population at risk. The prevalence of epilepsy amongst the parents, about 5%, is higher than most statistics for this condition would lead one to expect. The aetiological significance of this observation for hemiplegia is difficult to assess, but clearly a larger series of family studies might be rewarding.

Mental retardation was present in the parent or parents only in 5 cases, in a sibling or siblings only in 3, and in parents and siblings in 5. At least 13 of the 150 parents of children /

TABLE 16

Family History of Patients with Hemiplegia - Cerebral Palsy

Case No.	Mother	Father	Siblings		Stillbirths and Neonatal Deaths	Relatives
			Normal	Abnormal		
Congenital Hemiplegia.						
29	Feeble minded	Epileptic	1	-	-	M.G.M. diplegic
201	Healthy	Healthy	-	-	-	Mother's sister's child diplegic
39	Healthy	Alcoholic	1	1 diplegic	1 infant death (congenital heart)	-
Hemiplegia of Unknown Origin.						
199	Feeble minded	Healthy	1	1 diplegic (Case 98) 1 mentally defective	-	-

TABLE 17.

Family History of Patients with Hemiplegia - Epilepsy

Case No.	Mother	Father	Siblings		Stillbirths and Neonatal Deaths	Relatives
			Normal	Abnormal		
Congenital Hemiplegia.						
29	Feeble minded	Epileptic	1	-	-	M.G.M. diplegic
100	Healthy	Epileptic	2	-	-	2 paternal uncles epileptic
192	Epileptic	Healthy	1	-	-	-
Hemiplegia of Unknown Origin.						
134	Healthy	Healthy	10	1 epileptic	1 neonatal death	-
Acquired Hemiplegia.						
93	Healthy	Healthy	2	1 epileptic	-	-
19	Epileptic	Healthy	12	-	1 stillbirth 1 neonatal death	1 maternal aunt epileptic
48	Epileptic	Healthy	1	-	-	-
52	Epileptic	Healthy	-	-	1 stillbirth	-
132	Psychopathic	Epileptic	2	-	-	Paternal uncle and grandfather epileptic

TABLE 18.

Family History of Patients with Hemiplegia - Mental Retardation

Case No.	Mother	Father	Siblings Normal	Siblings Abnormal	Stillbirths and Neonatal Deaths	Relatives
Congenital Cases.						
109	Poor intelligence	Poor intelligence	3	1 mentally retarded	1 stillbirth	Maternal uncle and his son mentally retarded
5	Feeble minded	Epileptic	0	1 mentally defective	1 stillbirth	-
162	Acquired hemiplegia	Healthy	3	2 mentally retarded	-	-
170	Feeble minded	Healthy	5	2 mentally retarded	-	-
111	Feeble minded	Healthy	0	-	-	-
Hemiplegia of Unknown Origin.						
199	Feeble minded	Healthy	1	1 diplegic 1 feeble minded	-	-
58	Feeble minded	Healthy	3	-	-	-
168	Feeble minded	Feeble minded	0	-	-	-
Acquired Hemiplegia.						
19	Feeble minded	Healthy	11	-	Twins - 1 stillborn 1 neonatal death	Maternal uncle died of convulsions
50	Healthy	Alcoholic	3	1 "congenital chorea" and feeble minded	-	-
89	Feeble minded	Feeble minded	1	2 "psychopathic" 1 feeble minded	-	-
188	Feeble minded	Feeble minded	6	1 feeble minded	1 stillbirth	-
184	Feeble minded Alcoholic	Healthy	2	-	-	-

children suffering from hemiplegia showed significant retardation, giving a prevalence of 8.6%. The great majority of these parents were in Social Classes IV and V, some in very poor social conditions, as has been described. The exposure to infection suffered by young children in these circumstances, together with lack of intelligent care, might be expected to predispose to the occurrence of acquired hemiplegia. On the other hand it is difficult to explain the excess of children with congenital hemiplegia born to mentally retarded parents so simply. Possibly there is some inherited predisposition for the brain to suffer permanent damage more easily in these cases. The truth of such a speculation would appear to be almost impossible to prove or disprove without an investigation on a national scale.

Maternal Factors.

Ages of Mothers. The distribution of hemiplegic patients by the age of their mothers at the time of birth is shown in Table 19 . It is compared to the distribution of all live births in Counties of Cities given by the Registrar General for Scotland (1952). When the whole group of hemiplegic patients is considered it is found that there is no significant statistical difference in the distribution, though it has to be noted that the series of hemiplegic patients is relatively small.

On the other hand, when congenital cases are considered, it is found that 5 of the 30 cases (approximately 17%) had mothers aged over 40 years at the time of birth. In spite of the fact that there were more first-born children in the congenital /

TABLE 19.

Distribution of Cases of Hemiplegia by Maternal Age Compared to Distribution of Live Births in Counties of Cities (Registrar General, 1951)

Maternal Age	R.G. %	Number of Cases of Hemiplegia				Approx.%
		Congenital	Unknown	Acquired	Total	
Under 20	4.08	0	0	0	0	0
20 - 24	27.22	9	3	15	27	36
25 - 29	31.31	11	2	9	22	28
30 - 34	21.01	5	2	5	12	16
35 - 39	12.40	0	2	3	5	7.5
40 - 44	3.56	4	1	0	5	7.5
45 - 49	0.23	1	0	0	1	1.0
50 +	0.00	0	0	0	0	0
Unknown	0	0	2	1	3	4
T o t a l	100 %	30	12	33	75	100 %

TABLE 20.

The Average Age of Mothers
At the Time of Birth of Hemiplegic Children

	1st Pregnancy	2nd Pregnancy	3rd Pregnancy	4th and Subsequent	Unknown	All Pregnancies
Congenital Hemiplegia						
No. of cases	15	5	5	5	0	30
Average age of mothers	26.9	23.2	32.0	37.4	-	28.9
Acquired Hemiplegia						
No. of cases	12	9	4	7	1	33
Average age of mothers	23.7	25.8	25.0	33.3	-	26.5

congenital than the acquired group, there is a significantly higher proportion of patients with mothers over 40 in the former as compared to the latter (using the X^2 test). The difference in the proportion of mothers over the age of 40 in the congenital hemiplegic group and in the series of the Registrar General is also significant. P less than .01

The average ages of mothers are compared by the place of the child in the family in congenital and acquired hemiplegia in Table 20. It will be observed that the average age of the mother at the time of the delivery of a first child suffering from hemiplegia is some three years greater than for the first child with acquired hemiplegia, and is greater than the average age of mothers whose second children were hemiplegic. The average age of mothers rises much more sharply for each gestation in congenital hemiplegia than it does in acquired hemiplegia.

Maternal Health. The neurological, mental and psychiatric abnormalities of the mothers of hemiplegic children have already been considered, but physical illnesses which might, in particular, explain the lower fertility of mothers of congenital cases have not been described.

In Table 21. are shown the chronic physical illnesses which were not apparent complications of the pregnancies which resulted in the birth of the affected patients.

It will be seen that three mothers in the group of children suffering from acquired hemiplegia and one in the group with hemiplegia of unknown origin had tuberculosis, a reflection of the /

TABLE 21.

Health of Mothers of Hemiplegic Patients

Case No.	Birth Weight lbs.	ozs.	Maternal Age	Health of Mother	Health of Father	Social Class	Obstetric History	Siblings
Congenital Hemiplegia.								
162	4	9	42	Severe anaemia Hemiplegia	Good	V	4 abnormal deliveries 1 normal labour	Healthy
180	10	4½	21	Good - but pre-diabetic	Good	III	2 normal deliveries	Healthy
Hemiplegia of Unknown Origin.								
	6	3¾	24	Bronchitis and asthma	Good	V	2 normal deliveries 1 assisted breech 1 forceps	Healthy
160	Unknown		?	Pulmonary tuberculosis	Good	III	Unknown	Unknown
Acquired Hemiplegia.								
110	7	8	22	Pulmonary tuberculosis	Good	III	1 normal delivery 1 miscarriage	1 healthy
132	9	8	28	Pulmonary tuberculosis Psychopathic	Epileptic	V	2 normal deliveries	2 healthy
195	7	4	23	Pulmonary tuberculosis	Pulmonary tuberculosis	V	6 normal deliveries	1 epileptic 5 normal
164	5	10½	27	Thyrotoxicosis	Good	V	2 normal deliveries	2 healthy

TABLE 22.

Number of Pregnancies and Average Number of Pregnancies per Mother by Origin of Hemiplegia (Including Pregnancies Resulting in Birth of Patients)

Number of Cases		Number of Pregnancies	Average per Mother
Congenital	30	84	2.8
Unknown Aetiology	11	42	3.8
Acquired	33	134	4
All	74	260	3.5

the social conditions in which they lived rather than an aetiological factor of direct importance in hemiplegia. One mother in the group of hemiplegia of unknown origin (Case 36) had bronchitis and asthma. The mother in Case 180 in the congenital group was a pre-diabetic, suspected of being so on the basis of the increasing birth weights of successive children and confirmed by later investigations. Case 162 suffered from severe anaemia and hemiplegia. In Case 164 of the acquired group the mother was thyrotoxic.

The Fertility of Mothers of Hemiplegic Patients. The total number of pregnancies to mothers of hemiplegic patients were studied. There was a total of 260 of these distributed as shown in Table 22 . It will be seen that mothers of children with congenital hemiplegia have fewer pregnancies than the mothers with children suffering from acquired hemiplegia or hemiplegia of unknown origin. As has been noted, the number of pregnancies per mother is greater in the lower compared to the upper social classes.

To some extent the apparently higher fertility in the acquired group may be the result of the fact that many acquired cases of hemiplegia are the result of complications of contagious diseases occurring at an early age. These are more likely to occur in the young children of big families, where older children are liable to bring infections into the household, than in small families. But there may also be a reduced fertility in the mothers of congenital cases.

The Spacing of Pregnancies. The period elapsing between the /

TABLE 23.

Spacing of Pregnancies to Mothers of Hemiplegic Children in Years Between Dates of Delivery or Abortion

	Origin of Hemiplegia		
	Congenital	Unknown	Acquired
Number of previous pregnancies considered	36	19	63
Number of subsequent pregnancies	18	12	38
Average number of: Years between previous deliveries	2	1.7	2.3
Average number of: Years between previous child and patient	2.8	2.1	3.0
Average number of: Years between patient and subsequent pregnancy	3.2	2.5	3.1
Average number of: Years between subsequent pregnancies	2.3	1.3	1.7

the dates of abortions or the delivery of still- and live-born children was estimated, and the average number of years between these are shown in Table 23 . It will be seen that the average period elapsing between the pregnancies of the mothers of congenital hemiplegic patients is not significantly different from that of the mothers of patients suffering from acquired hemiplegia.

It is very difficult to see why there should be a considerably longer period, on the average, between the previous pregnancy and the pregnancy with the patient in all three groups of hemiplegic patients. It might be assumed that in the congenital cases there is some temporary infertility of the mother which might also be expressed in the creation of an abnormal foetus - the hemiplegic patient. But this possible explanation appears to be unlikely to apply to the mothers in the acquired group.

The most immediately presenting possible explanation for the longer period between the birth of the patient and the subsequent pregnancy than between either previous or later pregnancies would appear to be that the parents were deliberately practising contraception following the birth of an abnormal child or of a child becoming abnormal in the early years of life. A few parents did admit to practising contraception in an attempt to prevent the arrival of other children until the affected child should be old enough to demand less constant attention, but their numbers were few.

Foetal and Infant Loss to Mothers of Hemiplegic Children.

The obstetric histories of the mothers of hemiplegic patients are of some interest from the point of view of determining to what extent the liability of the mothers of children with congenital hemiplegia to have other abnormal pregnancies differs from that of those with children suffering from acquired hemiplegia. As has already been indicated, there is no significant difference in the proportion of abnormal siblings in these groups, but the proportions of abnormal other pregnancies, labours or deliveries is higher in the group of congenital hemiplegia than in acquired hemiplegia (Table 24).

Though the total foetal and infant loss is not significantly different between the groups of congenital and acquired hemiplegia (approximately 21% and 17%), the way in which the loss is distributed is different. It will be observed that approximately 10% of other pregnancies to mothers of children with acquired hemiplegia ended in abortion, but that only approximately 4% of other pregnancies to the mothers of children with congenital hemiplegia ended in this way. On the other hand approximately 13% of the other pregnancies in the congenital group ended in stillbirth, but only 3% of those in the acquired group. Neither of these differences is of statistical significance (p = approximately 0.1) but the contrast in the groups is none the less of interest.

It will be seen from Table 25. that none of the pregnancies and deliveries of stillborn infants was normal, and that the stillbirths appear to be attributable to the effects of abnormal parturition. It seems possible therefore that the /

TABLE 24.

The Outcome of Pregnancies to the Mothers of Patients Suffering from Hemiplegia

Origin of Hemiplegia	Congenital	Acquired	Unknown	
Number of cases	30	33	11	
Total number of pregnancies	84	134	42	
Number resulting in birth of patients	30	33	11	
Number of other pregnancies	54	101	31	
Number of abortions	2	10	3	
Number of stillbirths	7	3	0	
Number of infant deaths	2	4	2	
Number of abnormal siblings	8	8	4	
Number of normal siblings	35	76	22	
Total number of siblings	43	84	26	
Abnormal other pregnancies	23	19	7	
Normal other pregnancies	31	82	24	
Stillbirth rate per 1000 other pregnancies	135	33	0	All 59
Infant mortality	44	45	72	50

TABLE 25.

Stillbirths to the Mothers of Patients Suffering from Hemiplegia

Case No.	Social Class	Maternal Age	Maternal Health	Number of Pregnancy Total Pregnancies	Pregnancy	Labour and Delivery	Maturity	Other Obstetric History
Congenital Cases.								
5	V	38	Good	2 / 4	Normal.	Premature spontaneous vertex delivery at 7½ months.	Premature.	1 forceps delivery. 1 normal pregnancy. 1 spontaneous vertex delivery after labour of 2 hours (patient).
70	V	45	Good	2 / 4	Antepartum haemorrhage. Prolonged labour.	Spontaneous vertex delivery.	Mature.	1 normal pregnancy and delivery.
				3 / 4	Antepartum haemorrhage. Prolonged labour.	Forceps delivery.	Mature.	1 prolonged labour and forceps delivery (patient).
80	IV	29	Good (Rh-ve)	1 / 3	Healthy.	Prolonged second stage of labour.	Mature.	1 normal pregnancy and delivery. 1 foetal distress (patient).
109	V	27	Low intelligence	5 / 6	Antepartum haemorrhage.	Premature spontaneous vertex delivery.	Premature.	4 normal. 1 threatened abortion and premature delivery (patient).
171	III	24	Healthy	4 / 5	Rhesus incompatibility.	Normal.	Mature.	1 normal.
				5 / 5	Rhesus incompatibility.	Normal.	Mature.	1 (patient) face to pubis, spontaneous vertex delivery.
Acquired Cases.								
19	III	31	Healthy	11 / 14 (t)	Healthy twin pregnancy.	Premature spontaneous vertex deliver[illegible]	Premature.	Twin stillborn. Other pregnancies and deliveries normal.
52	IV	23	Healthy	2 / 4	Healthy.	Premature spontaneou vertex delivery.	33 weeks	2 miscarriages. 1 normal (patient).
188	IV	34	Poor intelligence	2 / 9	Pre-eclampsia.	Breech extraction.	Postmature	1 forceps delivery. 1 miscarriage. 5 normal deliveries. 1 hydramnios and anaemia. Foetal distress (patient).

TABLE 26.

Infant Deaths to Mothers of Patients Suffering from Hemiplegia

Case No.	Social Class	Maternal Age	Maternal Health	Number of Pregnancy Total Pregnancies	Pregnancy	Labour and Delivery	Maturity	Cause of
Congenital Cases.								
39	III	29	Healthy	4 / 4	Healthy	Normal	Mature	Endocardi fibroela
84	III	24	Healthy	1 (t) / 3	Twin	Twin pregnancy	Premature	"Prematu (Twin hea
Acquired Cases.								
19	III	31	Healthy	6 / 14	Normal	Normal	Mature	Whooping at 3 wee
				11 (t) / 14	Twin	Spontaneous vertex delivery	Premature	Prematuri
165	III	25	Healthy	2 / 3	Healthy	Normal	Mature	Bilateral otitis media. Acute pancreatitis
169	IV	24	Healthy	5 (t) / 5	Healthy twin	Spontaneous vertex delivery	Premature	Spina bifida Multiple Died 13 days old (Twin well)
Unknown Origin.								
134	III	41	Healthy	4 / 12	Healthy	Normal	Mature	Convulsions (? meningitis) just before 1 year old
				10 (t) / 12	Twin	Assisted breech	Premature	"Prematurity and atelectasis" (Twin healthy)

TABLE 27a.

Abortions, Stillbirths and Infant Deaths to Mothers of Patients with Congenital Hemiplegia

Case No.	Social Class	Number of Pregnancies	Number of Abortions	Number of Stillbirths	Neonatal Deaths	Post-neonatal Deaths	Abnormal Siblings	Normal Siblings
5	V	4	0	1	0	0	0	2
39	III	4	0	0	0	1	1	1
70	V	4	0	2	0	0	0	1
80	IV	3	0	1	0	0	0	1
84	III	3	0	0	1 (t)	0	0	2
108	IV	3	1	0	0	0	0	1
109	V	6	0	1	0	0	1	3
162	V	7	1	0	0	0	0	5
171	III	5	0	2	0	0	0	2

TABLE 27b.

Abortions, Stillbirths and Infant Deaths to Mothers of Patients with Acquired Hemiplegia

Case No.	Social Class	Number of Pregnancies	Number of Abortions	Number of Stillbirths	Neonatal Deaths	Post-neonatal Deaths	Abnormal Siblings	Normal Siblings
17	III	5	1	0	0	0	0	2
19	III	14	0	1	1	0	0	12
50	V	6	1	0	0	0	1	3
52	IV	4	2	1	0	0	0	0
89	IV	8	2	0	0	0	1	4
110	IV	3	1	0	0	0	0	1
117	III	2	1	0	0	0	0	0
165	III	3	0	0	0	1	0	1
169	IV	6	0	0	0	1	2	3 (t)
188	IV	9	1	1	0	0	0	6
195	IV	9	1	0	0	0	1	6

TABLE 28.

Outcome of Other Pregnancies to Mothers of Patients with Hemiplegia

Origin of Hemiplegia	Numbers			Approximate Percentages		
	Congenital	Unknown	Acquired	Congenital	Unknown	Acquired
Total other pregnancies	54	31	101	100	100	100
Abortions	2	3	10	4	9	10
Stillbirths	7	0	3	13	0	3
Infant Deaths	2	2	4	4	8	4
Abnormal siblings	8	4	8	15	12	8
Normal siblings	35	22	76	64	71	75
Total siblings	43	26	84	80	84	83

the higher proportion of stillbirths occurring to mothers of congenital than of acquired cases may be a result of the greater frequency of abnormal parturition in the former.

The cases in which other pregnancies resulted in the birth of children dying in the first year of life are summarised in Table 26, 27. It will be seen that in three cases (Cases 84, 19 and 134) death was attributed to prematurity, and that all four infants were twins. In Case 169 there was a congenital abnormality and in Case 134 endocardial fibroelastosis. The other two deaths were from infection. These causes of death, prematurity, infection and congenital abnormality, cannot be considered to be any different in prevalence from those in the general population.

In Table 28. are shown the stillbirth and infant mortality rates expressed as the number in every ~~thousand~~ 100 other pregnancies. It will be seen that the rates are 59 and 50 respectively, approximately twice those prevalent in the general population. It would be of great interest to be able to analyse in detail the extent to which these deaths were the result of abnormalities of parturition similar to those encountered in the pregnancies and deliveries of hemiplegic patients.

Abnormal Children. Abnormal siblings of hemiplegic patients have already been described. It was pointed out that though there was a tendency for there to be more abnormal siblings in the lower social classes than the upper (largely owing to the higher proportion of mentally retarded children in /

in the former) there was no difference in the proportions in the three groups of congenital, acquired hemiplegia and hemiplegia of doubtful origin.

They were not, at that time, considered from the point of view of the failure which they represented on the part of the mother to produce healthy children. In Table 28 they are added to the numbers of infant deaths, stillbirths and abortions to give some picture of the efficiency of these mothers so far as bearing healthy children in other pregnancies is concerned. From this it will be observed that the proportion of pregnancies which result in the birth of normal children is only 64% in the group of congenital hemiplegia, and only 75% in the acquired hemiplegia group.

When only other pregnancies are considered, there is no statistically significant difference in the proportions of pregnancies resulting in the birth of healthy children in the congenital and acquired groups, but it will be observed that as many as a quarter of the other pregnancies in the congenital group resulted in foetal or neonatal death or in the birth of abnormal siblings. The stillbirth rate is, in fact, 135 per 1000 live and stillbirths in this group, and the infant mortality in the region of 44 per 1000.

It must be acknowledged, therefore, that the mothers of hemiplegic patients are inefficient so far as producing healthy children is concerned, and that not only is the foetal and infant mortality high, but the proportion of abnormal siblings is also large. In the congenital group, moreover, there is relatively lower fertility than in either the general population or in the group of mothers of patients with acquired hemiplegia.

The Patient.

Sex. The distribution by sex of hemiplegic patients is shown in Table(below) It will be seen that there is no significant difference in the ratio of males to females in acquired and congenital hemiplegia. It appears that the male has some predisposition to be affected by hemiplegia in childhood of congenital or acquired aetiology, as is also indicated by the similar findings of Asher and Schonell (1950) and Sachs and Peterson (1890).

Sex distribution of patients with hemiplegia by aetiology.

Aetiology	Male	Female	Total
Congenital	20	10	30
Unknown	9	3	12
Acquired	23	10	33
All aetiologies	52	23	75

This finding contradicts a number of earlier reviews in which an apparently equal sex incidence was found (Lovett, 1888; Osler, 1889; Freud and Rie, 1891).

Place in Family.

In Table 29 is shown the distribution of hemiplegic patients by their place in the family compared to the distribution of live births in Counties of Cities as presented /

TABLE 29

Place in Family
of hemiplegic patients

Number of Pregnancy	Congenital	Unknown	Acquired	All	%	R.G.%
1	15	4	12	31	41	35.02
2	5	4	9	18	24	28.76
3	5	0	4	9	12	16.46
4	2	1	1	4	5	8.50
5	1	0	4	5	7	4.49
6	2	0	1	3	4	2.64
7	0	0	1	1	1	1.52
8 +	0	1	1	2	3	2.45
Unknown	0	2	0	2	3	0.16
Total	30	12	33	75	100	100.00

Illegitimate.

Congenital	Unknown	Acquired
2	0	2

TABLE 30.

Distribution of Patients with Congenital Hemiplegia by Place in the Family and Expected Distribution, Taking into Consideration Family Size

Place in Family	Observed	Expected
1	15	11
2	5	8
3	5	6
4	2	2
5	1	1
6 or later	2	2

presented by the Registrar General (1952). The distribution of all hemiplegic patients is not significantly different from the latter, but when the hemiplegic group is broken down by aetiology it is seen that there is an apparent excess of first-born children in the congenital group. This excess is of probable statistical significance and is in keeping with the frequently stated opinion that hemiplegia is commoner in first-born children than in the later-born.

Further examination of the families shows that most of them are very small, however, and when, as in Table 30 , a comparison is made between the observed and expected distribution of patients by place in the family it is seen that there is no significant difference. In fact, congenital hemiplegic patients are no more often first-born than would be expected when the small size of the families of their mothers is considered.

The Aetiological Significance of the Birth Weight.

The birth weights were known in 71 of the 75 patients suffering from hemiplegia. When the birth weights of all the patients are considered it is found that they do not differ significantly from the figures obtained in an unselected series of consecutive hospital births by Drillien (1947). These latter figures are obtained from a study of women admitted to an Edinburgh maternity hospital. They have the advantage of being based on a population drawn very largely from a district similar to that covered by the survey. They have the disadvantage /

TABLE 31.

Distribution of cases of hemiplegia by birth weight compared to a series of hospital births. (Drillien, 1947)

Birth Weight	Congenital	Approx. %	Hemiplegia Unknown aet.	Acquired	Approx. %	Total	Approx. %	Drillien. Edinburgh Hosp. Births	Approx. %
3.1 - 3.8	1	3	0 -	0	0	1	1)	7	0.1
							)	21	0.3
3.9 - 4.0	0	0	0 -	1	3	1	1	48	0.7
4.1 - 4.8	0	0	0 -	0	0	0	0	91	1.3
4.9 - 5.0	2	7	0 -	1	3	3	4	108	1.5
5.1 - 5.8	1	3	0 -	0	0	1	1	193	2.8
5.9 - 6.0	1	3	0 -	2	6	3	4	421	6.0
6.1 - 6.8	5	17	3 -	1	3	9	12	909	13.0
6.9 - 7.0	0	0	3 -	3	9	6	8	1139	16.3
7.1 - 7.8	7	23	1 -	12	37	20	27	1313	14.5
7.9 - 8.0	1	3	0 -	2	6	3	4	1189	15.6
8.1 - 8.8	6	20	0 -	2	6	8	12	681	9.7
8.9 - 9.0	2	7	2 -	5	15	9	12	512	7.7
9.1 - 9.8	2	7	1 -	0	0	3	4	260	3.7
9.9 - 10.0	1	3	0 -	2	6	3	4	200	2.7
10.1 +	1	3	0 -	0	0	1	1	30	4.3
Unknown	0	0	2 -	2	6	4	5	0	0
Totals	30	99.0	12 -	33	100	75	100	7122	100.2

disadvantage of including a higher proportion of women showing abnormalities of pregnancy and delivery than might be expected in a sample of the population at large. The figures used for comparison are those for the numbers of surviving infants of each birth weight, including multiple births, of 4693 children of primiparous and 2329 multiparous women. This ratio of approximately two primiparous to every multiparous woman is in contrast to the ratio of approximately two primiparous to every three multiparous mothers of hemiplegic children encountered in the survey. These differences are not reflected by marked differences in the survival of babies of similar birth weights, however, and the figures give some indication of the distribution of babies by birth weight in the Edinburgh population. The most marked differences are probably that the proportion of premature babies and the proportion of unusually heavy babies is slightly greater in the hospital series than in the population at risk.

When the distribution of hemiplegic patients by birth weight is compared to the hospital series it is found that the differences are remarkably slight. On breaking down the hemiplegic group into patients suffering from congenital and acquired hemiplegia, however, it is found that whereas the distribution for the acquired group is very similar to the hospital series, that for congenital hemiplegia is significantly different. The proportion of babies with birth weights 5lbs. 8 ozs. and under 9 lbs., 73%, may be compared to the significantly higher proportion for acquired hemiplegia, 82%, or for the hospital series, 82.8%. On the other hand the

proportion /

proportion of congenital hemiplegic patients with birth weights of 5 lbs. 8 ozs. or under, 13%, was significantly higher than for the acquired group or the hospital series, 6% and 6.7% respectively. The proportion with birth weights over 9 lbs. was significantly higher than that in the acquired group, and higher, but not significantly higher statistically, than that in the hospital series.

On the basis of these figures it may be stated that there appears to be no marked predisposition of children of low or high birth weights to develop acquired hemiplegia. There appears to be some relationship, however, between the occurrence of hemiplegia and a low or high birth weight in congenital hemiplegia. The possible nature of the relationship will be discussed when the aetiology of congenital hemiplegia is considered in more detail.

CHAPTER 4b

The Birth Histories of Patients Suffering From Congenital Hemiplegia.

In order to simplify description, cases have been divided into four groups according to whether disorders of pregnancy or delivery or both or neither were present. It will be seen from Tables 32 to 35 that one patient had apparently normal pregnancy and delivery, six had abnormal pregnancy but normal delivery, seven had normal pregnancy but abnormal delivery, and sixteen had abnormal pregnancy and delivery. Thus in over half the cases abnormalities of both pregnancy and delivery were present. Twenty-four of the patients were noted /

TABLE 32.

THE BIRTH HISTORIES OF PATIENTS SUFFERING FROM CONGENITAL HEMIPLEGIA

Group 1. No Apparent Abnormality of Pregnancy, Labour and Delivery

Case No.	Social Class	Number of Pregnancy	Maternal Age	Pregnancy	Duration of Labour (hours)	Mode of Delivery	Drugs	Birth Weight lb.oz.	Estimated Gestation (weeks)	Neonatal Period	Placenta
84	III	2	24	Normal	17	Spontaneous vertex	None	9 -	40	Apnoea for 10 minutes. Huge caput. Cyanosis for 7 days.	Unknown

TABLE 33.

THE BIRTH HISTORIES OF PATIENTS SUFFERING FROM CONGENITAL HEMIPLEGIA

Group 2. Abnormal Pregnancy, Normal Labour and Delivery

Case No.	Social Class	Number of Pregnancy	Maternal Age	Abnormality of Pregnancy	Duration of Labour (hours)	Mode of Delivery	Drugs	Estimated Gestation (weeks)	Birth Weight lb.oz.	Neonatal Period	Placenta
5	V	3	40	Antepartum haemorrhage at 35 weeks	2 +	Spontaneous vertex	None	35	4 12	Apnoeic	Unknown
43	IV	2	23	Moderately severe pre-eclampsia	4	Spontaneous vertex	Chloroform	40	7 -	Apnoeic for some minutes	Unknown
67	III	3	29	Severe pre-eclampsia	22	Spontaneous vertex	Chloroform	40	8 1½	Apnoeic for 20 minutes. Fits 3 days. "Cerebral" 7 days.	1 lb. 8 oz. Many infarcts
109	IV	6	27	Threatened abortion at one and 3 months. Hyperemesis. Premature onset of labour.	2 - 3	Premature spontaneous vertex	None	28	3 2	Very feeble. Apnoeic for minutes. Incubator 3 weeks.	Unknown
171	III	1	29	Moderately severe pre-eclampsia.	1	Spontaneous vertex	Minnit's apparatus	43	7 3½	Severe apnoea at birth. Slow to gain weight.	1 lb. 4 oz.
201	III	1	30	Pre-eclampsia. Antepartum haemorrhage at 40 weeks.	3½	Spontaneous vertex	Minnit's apparatus	40	6 2½	Some minutes delay in breathing. Large caput.	Many infarcts

TABLE 34.

THE BIRTH HISTORIES OF PATIENTS SUFFERING FROM CONGENITAL HEMIPLEGIA

Group 3. Normal Pregnancy, Abnormal Labour or Delivery

Case No.	Social Class	Number of Pregnancy	Maternal Age	Pregnancy	Abnormality of Labour or Delivery	Duration of Labour (hours)	Drugs	Estimated Gestation (weeks)	Birth Weight lb.oz.	Neonatal Period	Placenta
29	IV	2	22	Normal	Foetal distress. Spontaneous vertex delivery with cord round neck.	7	Chloroform	36	5 8	Apnoea 15 minutes. "Cerebral" 3 days.	Unknown
39	III	3	31	Normal	Prolonged labour. Spontaneous vertex delivery.	120	None	43	7 -	Well	Unknown
71	IV	1	23	Normal	Rigid soft parts. Foetal distress. Difficult high forceps delivery.	57	General anaesthesia	40	7 1¼	Huge caput. Apnoeic for 20 minutes. "Cerebral" 14 days.	Unknown
97	III	4	40	Normal	Arrest due to outlet contraction. Low forceps delivery.	9	Chloroform	40	8 3	No abnormality.	Unknown
107	II	1	28	Normal	"Dystonia". Breech extraction.	7	General anaesthesia	40	8 -	Apnoea. Fits and "cerebral" for 5 days.	Unknown
180	III	1	21	Normal	Prolonged second stage labour. Low forceps delivery for outlet contraction.	22	Chloroform	40	8 4	"Some asphyxia". Fits for 4 days.	Many infarcts
192	V	1	28	Normal	Very prolonged labour. Spontaneous vertex delivery.	120	Morphia and chloroform	43	8 12	"Some apnoea". "Slow to revive".	Unknown

noted to be apnoeic for a period following birth, and two were jaundiced. Only five children were apparently normal immediately after birth and all showed signs of nervous disorder within the first ten days of life.

In Tables 32 to 35 are summarised the case histories of the thirty patients with congenital hemiplegia. Each table will be considered briefly in turn.

Table 32. No apparent abnormality of pregnancy or delivery.

There was one case in this category. Case 84 was delivered at home by a midwife from a maternity hospital. The previous pregnancy had resulted in twins being born, the second of which, delivered by the breech, died. The patient was delivered spontaneously after a labour of 17 hours. The mother described it as being more exhausting than her first, with very severe pains for which she was given Minnitt's apparatus to use. The infant was delivered by the vertex and was blue and apnoeic for ten minutes at birth, with a huge caput. The cyanosis persisted for one week, but the child fed normally and was active except that the parents noted he used his left arm more than his right. On examination at the age of 6 years a mild right hemiplegia with rather marked trophic changes was found. The formal history in this case cannot be considered as indicating definite abnormality of pregnancy or delivery, but the fact that the child was large, weighing 9 pounds, and the mother small, together with the history of apnoea, cyanosis and a "huge caput" makes it probable that the labour was a severe one from the infant's

point /

point of view as well as the mother's.

Table 33. Abnormal pregnancy, apparently normal delivery.

Six children had histories of abnormal pregnancies followed by apparently normal spontaneous vertex deliveries. Case 109 had a history of the mother having very severe hyperemesis in the first three months of pregnancy and persistent intermittent vomiting thereafter. She had threatened abortions at six weeks and three months gestation, and felt ill throughout pregnancy. The child was born at home after a labour of between two and three hours, in a state of apnoea and at an estimated gestation of 28 weeks. The infant was rushed to hospital and incubated for three weeks.

In five cases the disorders of pregnancy were confined to the last trimester of pregnancy. Four mothers had pre-eclampsia. In Cases 43 and 176 the condition was moderately severe. In Case 176 the mother was admitted to hospital two weeks before term in a very oedematous condition but with only slight albuminuria and a blood pressure of 145/90. In Cases 67 and 201 the pre-eclamptic toxaemia was stated to be severe. In the latter case it was complicated by antepartum haemorrhage at the onset of labour and by postpartum eclampsia which required very heavy sedation. Both infants were spontaneous vertex delivieries, after 22 hours in Case 67 and after $3\frac{1}{2}$ hours in Case 201. Both infants were apnoeic following delivery.

Case 5 was delivered at home in very poor social conditions after a premature labour lasting a few minutes over two hours, which /

which followed a moderately severe antepartum haemorrhage. The child weighed $4\frac{3}{4}$ lbs. and was apnoeic for some minutes. The estimated gestation was 35 weeks. On examination at the age of 15 the patient showed evidence of a lesion involving the left temporal lobe.

Of the 6 patients with abnormal pregnancy and apparently normal delivery only 2 were first-born.

Table 34 . Apparently normal pregnancy, abnormal delivery.

Seven patients had histories suggesting that pregnancy had been normal and delivery complicated. Four were first-born children; only one was premature. Delivery was spontaneous by the vertex in 3 cases. In one of these (Case 29) foetal distress was noted during labour. This child was delivered after 7 hours with the cord round the neck. He was apnoeic following birth. He had a birth weight of $5\frac{1}{2}$ lbs. and was "cerebral" for 3 days. Of the other two cases who were spontaneous vertex deliveries both had prolonged second stage of labour, lasting 120 hours in Case 192, and pains were intermittent for 5 days in Case 39. Case 192 was apnoeic at birth; Case 39 appeared to have no ill effects. It is of interest that in Case 192, whose birth weight was 8 lbs. 12 ozs., the estimated gestation was 43 weeks. Three injections of morphia totalling three-quarters of a grain were given during the 120 hours of labour.

One patient (Case 107) was delivered by breech extraction under general anaesthetic after a labour of 7 hours. The extraction was a difficult one and the child was noted to have inspired /

inspired prior to delivery of the head. It was apnoeic on delivery and had convulsions for 5 days. The birth weight was 8 lbs.

Three patients were delivered by forceps. In Case 71 a difficult high forceps delivery was performed on account of foetal distress and high arrest of the foetal head after a labour of 57 hours. Unfortunately, though the delivery was in hospital the mother's maternity notes could not be traced. The child was apnoeic for 20 minutes and "cerebral" for 14 days. Case 97 was delivered by low forceps after a labour of 9 hours, on account of outlet contraction of the pelvis and failure to advance during the second stage of labour. The birth weight was 8 lbs. 3 ozs. The child breathed immediately on delivery. Case 180 was delivered by low forceps after a labour of 22 hours. Caput could be seen for half an hour before delivery. The child weighed 8 lbs. 4 ozs. and had apnoea for some minutes.

Table 35. Abnormal pregnancy and delivery.

Sixteen of the 30 patients were in this category, and 12 had apnoea of varying degrees of severity following birth. In 7 cases disorders of pregnancy occurred in the first 6 months of pregnancy as well as in the last trimester. Two patients had evidence of rhesus incompatibility (Cases 171 and 80). In the former case delivery was spontaneous by the vertex, but the mother was badly torn and the occipito posterior position was not corrected prior to delivery which was after a labour of 8 hours. The child weighed 8 lbs. 13 ozs. and /

and was jaundiced at birth. Transfusions of blood were given into the saggital sinus in the week following delivery. At the age of 10 years the child showed a severe hemiplegia, slight hemianaesthesia and focal epileptic fits with a pronounced sensory aura confined to the hemiplegic side. It seems possible that localised damage to the sensory cortex may have been caused at the time of the transfusion, but in view of the other history the case must be included in the congenital group.

In Case 80 maternal antibodies were noted at the 32nd week of pregnancy and the mother was admitted to hospital for her confinement. Labour occurred at term and lasted 12 hours. During it there was a marked outpouring of meconium, and the foetal heart was noted to be very variable in rate. Spontaneous delivery by the vertex occurred. The child weighed 6 lbs. 3½ ozs. and was jaundiced. There was apnoea for some minutes. Exchange transfusion was performed by the saphenous route since the umbilical vessels were found unsuitable. During transfusion the baby collapsed on two occasions, and following it the transfused lower limb showed evidence of extensive femoral vein thrombosis. Thus both the cases of rhesus incompatibility had complications of delivery and of transfusion.

Two patients had threatened or attempted abortion. In Case 70 pituitrin injections were given to induce labour at 41 weeks gestation and to encourage uterine contractions during it. After a labour of 13 hours during which morphia was also given, a high forceps delivery resulted in the birth of /

of a 9 lbs. male infant in a state of asphyxia pallida. In this case the mother had previously given birth to two very large stillbirths some weeks after the expected date of delivery. In Case 108 abortion was attempted at about four months' gestation. The mother was admitted to hospital three weeks prior to delivery on account of persistent anaemia. Her haemoglobin was never more than 55%. She had had a heavy fall at seven months resulting in much abdominal bruising and necessitating rest in bed for one week. External version was performed at 32 weeks. During labour there was evidence of foetal distress and after 19½ hours a female child weighing 7 lbs. 2ozs. was delivered with the cord around the neck and in a state of severe apnoea. Cyanosis persisted for one week.

In Case 167 there was a history of recurrent pyelitis before and during pregnancy, with intermittent fevers from the third month onwards. For the last 3 months of her pregnancy the mother was in hospital and received sulphonamides with little effect. She was given morphia, a total of half a grain during labour, because her pains were thought to be due to renal colic until shortly before delivery.

Six cases had a history of pre-eclamptic toxaemia during pregnancy. It was severe in three cases, being accompanied by marked hypertension and albuminuria (Cases 90, 123, 143). In all three labour was prolonged. The child was delivered at the second attempt by high forceps in Case 90 after heavy pethidine analgesia, ether anaesthesia and spinal anaesthesia had been given. Labour lasted 72½ hours and during it foetal distress was noted. The child was apnoeic for 15 minutes after /

after birth and had generalised convulsions for 7 days. In the other two cases foetal distress was present during the prolonged labours, attributed to rigid soft parts in Case 123 and to disproportion in Case 143. Delivery was spontaneous by the vertex in the former case and by mid cavity forceps in the latter. In Case 123 heroin and chloroform were given during labour and in Case 143 morphia and chloroform. Both infants were apnoeic after birth.

Three patients had moderately severe pre-eclamptic toxaemia followed by complicated labour. In Case 86 low forceps delivery was performed for outlet arrest after a labour of 10 hours. The infant breathed immediately on delivery. In Case 111 there was uterine inertia accompanied by foetal distress, the heart rate being 80 persistently, and much meconium being produced. Pentothal was given repeatedly and under cyclopropane anaesthesia mid cavity forceps was performed. The child was apnoeic for 10 minutes and very cyanosed thereafter. In Case 100 moderately severe pre-eclamptic toxaemia was complicated by the occurrence of profuse antepartum haemorrhage at the onset of labour. Haemorrhage continued during the 8 hours' labour and resulted in the spontaneous vertex delivery of a male infant weighing 6 lbs. 5 ozs. in a state of apnoea. It is of interest that in the six cases in which pre-eclamptic toxaemia occurred the placentae were noted to be infarcted in three cases and offensive in one.

Three patients had antepartum haemorrhage unassociated with toxaemia. In Cases 79 and 170 haemorrhage occurred immediately /

immediately prior to the onset of labour at an estimated gestation of 40 weeks. In Case 79 pains were rather poor and after 15 hours the attendant doctor gave her an injection of pituitrin. Very intense pains resulted. Foetal distress was diagnosed and the child was delivered with great difficulty by high forceps through the partially dilated cervix. Artificial respiration resulted in regular respiration only after one hour. In Case 170 delivery was spontaneous by the vertex after a labour of 8 hours. The infant showed evidence of foetal distress during labour, the pulse rate falling to a persistent rate of 70 per minute shortly before delivery, and there was copious outpouring of meconium. The child weighed 6 lbs. $\frac{3}{4}$ oz. and was apnoeic for some minutes. Antepartum haemorrhage occurred immediately before the onset of labour in Case 74. Labour was extremely rapid and lasted for less than 30 minutes. Pains were extremely severe. The child weighed 6 lbs. $3\frac{1}{2}$ ozs. and breathed at once.

The Neonatal Course of Patients with Congenital Hemiplegia.

Unfortunately it was in the study of the post-natal period that the notes of doctors, nursing homes and hospitals proved inadequate. It is very difficult to make any real assessment of the clinical condition of the children in the neonatal period on the basis of the limited information they provide in the majority of cases. Far too often comments on the state of the infant following birth are restricted to:- "Slow to breathe"; "Shocked but responded"; "Limp but responded to resuscitation". This is especially true of the earlier /

earlier cases in the series; there are much fuller clinical details of the patients born in hospital in the post-war period. Nevertheless the study of the neonatal period of the patients in the present series is rather limited and tentative because of the lack of reliable information.

The clinical details which are available are summarised in Table 36 . It will be seen that in 22 of the 30 patients there was a definite delay in the establishment of respiration. There appears to be no special correlation between the occurrence of disordered pregnancy or delivery or both periods with the incidence or severity of apnoea. Five of the 6 patients with abnormal pregnancies but apparently normal deliveries were apnoeic, and 11 of the 16 patients with abnormal pregnancies and deliveries. There appears to be no correlation between the occurrence of severity of apnoea and the severity of the hemiplegia eventually evident, nor did apnoeic infants appear to have ~~any~~ stormier neonatal courses than those without apnoea.

In 13 of the 22 apnoeic patients the exact duration of the apnoea is unknown. In 2 cases it was stated to have been 20 minutes, in 2 15 minutes, in 3 10 minutes. Three patients are said to have had a period of apnoea followed by irregular respiration for one hour in Case 79, two hours in Case 70, and 30 minutes in Case 167. One of the patients without apnoea and one with apnoea were jaundiced at birth due to rhesus incompatibility (Cases 80 and 171). One of the children without apnoea was born with two teeth (Case 86). Two of the remaining 4 infants who breathed at once after birth were noted

to /

to have a paretic upper limb within a few days of delivery (Cases 39 and 97). Both were subsequently found to be severely mentally defective.

Unfortunately details of the measures used to attempt to initiate respiration are poorly recorded. Details which are known are shown in the Table.(P 187).

Eight patients had generalised convulsions in the first month after delivery. In five cases the convulsions started on the day of birth, in two on the second day, and in one during sagittal sinus transfusion on the seventeenth day after delivery. Intermittent convulsions were present for periods varying between 2 days and 3 weeks. Of the eight patients with convulsions in the neonatal period four had epileptic fits in later childhood. This incidence of chronic epilepsy is no greater than for an unselected series of hemiplegic patients.

With the exception of Case 43 who was placid in the neonatal period, all the infants with convulsions were noted by their parents to be hyperexcitable, or "cerebral" by medical or nursing attendants. In addition to 7 of the 8 with convulsions, 5 of the 22 patients without convulsions in the neonatal period were also noted to be cerebral or restless or hyperexcitable. Frequently the parents noted that these children appeared to sleep little, wakened at the slightest sound and cried at night and in the day-time with great readiness. In 5 cases the children were specifically described by the mother as being "starey". Without exception /

exception the 12 patients in whom restlessness was apparent in the neonatal period had difficulty in feeding. The children were difficult to fix to the breast because, as one mother put it, "He didn't take the nipple smoothly and nicely, but took great jumps at it and kept missing, and when it did get near his mouth he didn't seem to know what to do with it." The difficulty appears to be due partly to the hypertonic state of the children, and their tendency to assume opisthotonic attitudes when stimulated, which inhibits the smooth working of the sucking reflex. In some cases a contributory factor was probably the jerking movements evident even when the child was not in a full manifest convulsion.

In addition to the difficulty in fixing to the breast or bottle, swallowing was impaired in some of the 12 hypertonic and overactive infants. This resulted in very slow feeding and a tendency to regurgitate milk very readily. The weight gain of 9 of the 12 hypertonic, overactive infants was stated to be "poor" in the first month of life, to be "fair" in two and "normal" in one. Of the 12 restless infants only 3 were breast fed.

The picture obtained from a study of the available descriptions by parents and from hospital notes of the restless jerking infant, starey-eyed and "too highly strung to feed" is very similar to that described as being typical of subdural haemorrhage by Craig (1938)

In contrast to the picture of the restless child is the picture of the "good baby", who spent most of his neonatal period sleeping, crying little, and frequently apparently too /

too weary to feed energetically or to finish his feed. The picture was very similar to that presented retrospectively by mothers of severely mentally defective children. These infants numbered 5 in the 30 with congenital hemiplegia. In 4 mental defect was present on examination in later childhood, and in 3 the hemiplegia was mild, mental defect being the striking finding (Cases 43, 97 and 7). The sluggish infants were also difficult to feed - "I had to pinch him all the time to keep him at it", said one mother. Fixing tended to be difficult, but the chief trouble was the slow speed at which the infants took their feed, their tendency to regurgitate and vomit, and their liability to fall asleep during it.

In 12 cases the behaviour of the infants was described as normal, and in one no information was forthcoming. Of the 12, feeding behaviour was stated to be normal in 9, and feeding was rather slow and difficult in 3 (Cases 74, 111, 100).

In 2 cases facial palsy was noted immediately following birth (Cases 71 and 143), and in 4 one upper limb was noted to be moved much less than the other in the first ten days of life (Cases 84, 192, 39 and 97) by medical attendants. It is unfortunately necessary to accept with great reserve the statements of relatives that the hemiplegic limbs were grossly contractured and deformed at birth, and that they noticed this as soon as they set eyes on the child. In the majority of cases, children with congenital hemiplegia who had not had fits or feeding disturbances were taken to their doctors only after 3 or 4 months had elapsed, and photographs taken during the neonatal period often failed to confirm the relatives' statements. /

TABLE 20

The neonatal course of patients with congenital hemiplegia

Group & Case No.	Abnormality of pregnancy.	Abnormality of pregnancy and labour	Bth.Wgt. lbs.oz.	Respiration	Other findings	Treatment	Onset of neonatal fits.	Duration of initial fits.	Behaviour	Feeding	Weight gain.	Final clinical picture
(1) 84	Normal.	Normal.	9 -	Apnoea 10 mins.	Huge caput. Cyanosed for 1 wk.	Artif.Resp. Oxygen.	-	-	Moved R. hand little. Otherwise well.	Normal.	Normal.	Mild right hemiplegia. Marked trophic changes.
(2) 5	Antepartum haem. at 35 weeks.	-	4 12	"Mins." apnoea.	Cyanosed.	Slapped. Coramine.	2nd.day	3 weeks.	Restless and cried much.	Slow with regurgit.	Slow.	Moderately severe left hemiplegia and epilepsy.
43	Mod. severe pre-eclampsia.	-	7 0	"Mins." apnoea.	Cyanosed.	Sedated phenobarbitone.	At birth	2 weeks.	Very still and placid.	Slow and difficult.	Poor.	Mild right hemiplegia.
67	Severe pre-eclampsia.	-	8 1½	20 mins. apnoea.	Opistalatonia and pale.	Artif.Resp. Oxygen.	First-day	3 weeks.	Irritable. Tense. Jumpy.	Slow.Some vomiting.	Poor.	Severe left hemiplegia. Epilepsy.
109	Threatened abort. Hyperemesis. Premature labour.	-	3 2	"Mins." apnoea.	Feeble. Premature.	Incubator for 3 wks.	-	-	"Seemed lazy".	Slow.	Fair.	Mod. severe right hemiplegia. Mental defect.
176	Mod. severe pre-eclampsia.	-	7 3½	Prolonged apnoea.	Post mature.	Lobeline into cord. Oxygen. Incubator.	-	-	Normal.	Normal.	Normal.	Severe left hemiplegia.
201	Sev. pre and post-partum eclampsia. Antepartum haem.	-	6 2½	"Mins." apnoea.	Huge caput. Much naevus.	Oxygen. Asp. of mucous.	-	-	Normal.	Normal.	Good.	Mild right hemiplegia.
(3) 29	Normal.	Foetal distress.Spont.del. Cord round neck.	5 8	Apnoea 15 mins.	Cyanosed.Irritable later.	Sedation.	Unknown	Unknown	Very restless.	Slow.	Fair.	Mild right hemiplegia. Mental defect. Epilepsy.
39	Normal.	Prolonged labour. Spont. vertex delivery.	7 -	No apnoea.	Well.	Nil.	-	-	Normal	Normal.	Good.	Severe right hemiplegia.
71	Normal.	Rigid soft parts.Foetal distress. Diff. high forceps delivery.	7 1¼	Apnoea 20 mins.	Huge caput. Left facial palsy.	Not recorded.	-	-	"Cerebral" and restless.	Normal.	Normal.	Mod. severe left hemiplegia.
97	Normal.	Prolonged 2nd. stage. Outlet contraction.	8 3	Normal.	R. arm flexed across chest.	-	-	-	Very sluggish and lazy.	Very slow.	Normal.	Severe right hemiplegia.
107	Normal.	Dystonia. Breech extraction.	8 0	"Mins." apnoea.	Unknown.	Sedated.	1st.day	5 days.	Irritable. Restless.	Difficult.	Slow.	Severe left hemiplegia. Epilepsy.
180	Normal.	Prolonged 2nd. stage. Low forceps delivery.	8 4	"Mins." apnoea.	-	Coramine into cord. Oxygen.	-	-	"Cerebral" and restless.	Very difficult.	Slow.	Severe left hemiplegia. Epilepsy.
192	Normal.	V. prolonged labour. Spont. vertex delivery.	8 12	"Mins." apnoea.	Flaccid right arm.	Slapped.	-	-	Irritable.	Slow. Difficult.	Unknown.	Severe right hemiplegia.
(4) 7	Fainting in last 2 weeks.	Dystonia. Mid cavity arrest. Mid cavity forceps.	9 13	Normal.	-	-	-	-	Very sluggish.	Diff.to fix. Regurgitation.	Slow.	Mild right hemiplegia. Mental defect.
70	Threatened abort. Pituitrin induction.	Pituitrin for uterine inertia. High forceps.	9 0	Irreg. 2 hours.	-	Artif.Resp. Injections.	1st.day	3 days	"Cerebral" for 5 days.	Very diff. Regurgitation.	Poor.	Mod. severe left hemiplegia.
74	Antepartum haem.	Precipitate labour.	6 3½	Normal.	-	-	-	-	Normal.	Slow.Would not suck.	Slow.	Mod. severe left hemiplegia.
79	Hydramnios.Antepartum haem.	Cervical dystonia.Pituitrin. High forceps delivery.	8 -¼	Irreg. 1 hour.	Cyanosed.	Hot baths. Coramine.	-	-	"Jumpy" and irritable for 1 week.	V. difficult to fix.	Poor.	Mild left hemiplegia.
80	Rhesus incompatibility.	Foetal distress. Spont. vertex delivery.	6 3½	"Mins." apnoea.	Jaundiced.	Exchange Transf.	-	-	Normal.	Normal.	Normal.	Mild right hemiplegia.
86	Pre-eclampsia.	Low forceps delivery for outlet arrest.	7 4	Normal.	2 teeth.	-	-	-	Very sluggish and lazy.	Slow.	Fair.	Severe right hemiplegia. Mental defect. Epilepsy.
90	Severe pre-eclampsia.	Uterine inertia. Failed forceps and high forceps delivery.	8 5	Apnoea 15 mins.	Persistent cyanosis.	Intubation. Oxygen.Coramine into cord.	1st. day	7 days	Very restless and irritable.	Difficult.	Poor.	Severe right hemiplegia.
100	Mod. sev. pre-eclampsia. Haem. at 39 weeks.	Intrapartum haem. Spont. vertex delivery.	6 5	Apnoea 10 mins.	Cyanosed.	Lobeline. Oxygen. Hot baths.	-	-	Normal.	Diff. to fix.Would not suck.	Poor.	Mild left hemiplegia. Mental defect.
108	Attemted abortion. Anaemia.Fall 7 mths. External version.	Foetal distress. Spont. vertex. Cord round neck.	7 2	"Mins." apnoea.	Persistent cyanosis.	Oxygen.Coramine.Incubator.	-	-	Stiff Right arm.	Normal.	Normal.	Severe right hemiplegia.
111	Moderately severe pre-eclampsia.	Uterine inertia.Foetal distress.Mid cavity forceps.	7 14½	Apnoea 10 mins.	V. cyanosed.	Oxygen.	-	-	Normal.	Slow.	Poor.	Severe right hemiplegia.
123	Sev.pre-eclampsia.	Rigid soft parts.Prolonged labour.Spont.vertex.deliv.	7 -	Bronchopneumonia.	V. cyanosed.	Lobeline. Coramine. Oxygen.	-	-	Normal.	Normal.	Normal.	Severe right hemiplegia.
143	Hyperemesis.Severe pre-eclampsia.	Prolonged labour.Foetal distress.Mid cavity forceps.	10 8½	Irreg.respir. 30 minutes.	Facial palsy. Cephalhaematoma.	Artif.resp. Oxygen.	-	-	Restless.Irritable. Cyanosed.	Diff. to fix.	Slow.	Mod. severe right hemiplegia.

	43	Mod. severe pre-eclampsia.						phenobar-bitone.			placid.	difficult.		
	67	Severe pre-eclampsia.	-	8	1½	20 mins. apnoea.	Opistalatonia and pale.	Artif.Resp. Oxygen.	First-day	3 weeks.	Irritable. Tense. Jumpy.	Slow.Some vomiting.	Poor.	Severe left hemiplegia. Epilepsy.
	109	Threatened abort. Hyperemesis. Premature labour.	-	3	2	"Mins." apnoea.	Feeble. Premature.	Incubator for 3 wks.	-	-	"Seemed lazy".	Slow.	Fair.	Mod. severe right hemiplegia. Mental defect.
	176	Mod. severe pre-eclampsia.	-	7	3½	Prolonged apnoea.	Post mature.	Lobeline into cord. Oxygen. Incubator.	-	-	Normal.	Normal.	Normal.	Severe left hemiplegia.
	201	Sev. pre and post-partum eclampsia. Antepartum haem.	-	6	2½	"Mins." apnoea.	Huge caput. Much naevus.	Oxygen. Asp. of mucous.	-	-	Normal.	Normal.	Good.	Mild right hemiplegia.
(3)	29	Normal.	Foetal distress.Spont.del. Cord round neck.	5	8	Apnoea 15 mins.	Cyanosed.Irritable later.	Sedation.	Unknown	Unknown	Very restless.	Slow.	Fair.	Mild right hemiplegia. Mental defect. Epilepsy.
	39	Normal.	Prolonged labour. Spont. vertex delivery.	7	-	No apnoea.	Well.	Nil.	-	-	Normal	Normal.	Good.	Severe right hemiplegia.
	71	Normal.	Rigid soft parts.Foetal distress. Diff. high forceps delivery.	7	1¼	Apnoea 20 mins.	Huge caput. Left facial palsy.	Not recorded.	-	-	"Cerebral" and restless.	Normal.	Normal.	Mod. severe left hemiplegia.
	97	Normal.	Prolonged 2nd. stage. Outlet contraction.	8	3	Normal.	R. arm flexed across chest.	-	-	-	Very sluggish and lazy.	Very slow.	Normal.	Severe right hemiplegia.
	107	Normal.	Dystonia. Breech extraction.	8	0	"Mins." apnoea.	Unknown.	Sedated.	1st.day	5 days.	Irritable. Restless.	Difficult.	Slow.	Severe left hemiplegia. Epilepsy.
	180	Normal.	Prolonged 2nd. stage. Low forceps delivery.	8	4	"Mins." apnoea.	-	Coramine into cord. Oxygen.	-	-	"Cerebral" and restless.	Very difficult.	Slow.	Severe left hemiplegia. Epilepsy.
	192	Normal.	V. prolonged labour. Spont. vertex delivery.	8	12	"Mins." apnoea.	Flaccid right arm.	Slapped.	-	-	Irritable.	Slow. Difficult.	Unknown.	Severe right hemiplegia.
(4)	7	Fainting in last 2 weeks.	Dystonia. Mid cavity arrest. Mid cavity forceps.	9	13	Normal.	-	-	-	-	Very sluggish.	Diff.to fix. Regurgitation.	Slow.	Mild right hemiplegia. Mental defect.
	70	Threatened abort. Pituitrin induction.	Pituitrin for uterine inertia. High forceps.	9	0	Irreg. 2 hours.	-	Artif.Resp. Injections.	1st.day	3 days	"Cerebral" for 5 days.	Very diff. Regurgitation.	Poor.	Mod. severe left hemiplegia.
	74	Antepartum haem.	Precipitate labour.	6	3½	Normal.	-	-	-	-	Normal.	Slow.Would not suck.	Slow.	Mod. severe left hemiplegia.
	79	Hydramnios.Antepartum haem.	Cervical dystonia.Pituitrin. High forceps delivery.	8	-¼	Irreg. 1 hour.	Cyanosed.	Hot baths. Coramine.	-	-	"Jumpy" and irritable for 1 week.	V. difficult to fix.	Poor.	Mild left hemiplegia.
	80	Rhesus incompatibility.	Foetal distress. Spont. vertex delivery.	6	3½	"Mins." apnoea.	Jaundiced.	Exchange Transf.	-	-	Normal.	Normal.	Normal.	Mild right hemiplegia.
	86	Pre-eclampsia.	Low forceps delivery for outlet arrest.	7	4	Normal.	2 teeth.	-	-	-	Very sluggish and lazy.	Slow.	Fair.	Severe right hemiplegia. Mental defect. Epilepsy.
	90	Severe pre-eclampsia.	Uterine inertia. Failed forceps and high forceps delivery.	8	5	Apnoea 15 mins.	Persistent cyanosis.	Intubation. Oxygen.Coramine into cord.	1st. day	7 days	Very restless and irritable.	Difficult.	Poor.	Severe right hemiplegia.
	100	Mod. sev. pre-eclampsia. Haem. at 39 weeks.	Intrapartum haem. Spont. vertex delivery.	6	5	Apnoea 10 mins.	Cyanosed.	Lobeline. Oxygen. Hot baths.	-	-	Normal.	Diff. to fix.Would not suck.	Poor.	Mild left hemiplegia. Mental defect.
	108	Attemted abortion. Anaemia.Fall 7 mths. External version.	Foetal distress. Spont. vertex. Cord round neck.	7	2	"Mins." apnoea.	Persistent cyanosis.	Oxygen.Coramine.Incubator.	-	-	Stiff Right arm.	Normal.	Normal.	Severe right hemiplegia.
	111	Moderately severe pre-eclampsia.	Uterine inertia.Foetal distress.Mid cavity forceps.	7	14½	Apnoea 10 mins.	V. cyanosed.	Oxygen.	-	-	Normal.	Slow.	Poor.	Severe right hemiplegia.
	123	Sev.pre-eclampsia.	Rigid soft parts.Prolonged labour.Spont.vertex.deliv.	7	-	Bronchopneumonia.	V. cyanosed.	Lobeline. Coramine. Oxygen.	-	-	Normal.	Normal.	Normal.	Severe right hemiplegia.
	143	Hyperemesis.Severe pre-eclampsia.	Prolonged labour.Foetal distress.Mid cavity forceps.	10	8½	Irreg.respir. 30 minutes.	Facial palsy. Cephalhaematoma.	Artif.resp. Oxygen.	-	-	Restless.Irritable. Cyanosed.	Diff. to fix.	Slow.	Mod. severe right hemiplegia.
	162	Twin pregnancy. Severe anaemia.	Prolonged 2nd.stage. Breech extraction.	4	9	Brief apnoea.	Premature. Feeble.	-	-	-	Unknown.	Unknown.	Unknown.	Mild right hemiplegia. Mental defect.
	167	Recurrent pyelitis.	Prolonged labour. Spont. vertex delivery.	5	4½	Apnoea few mins.	Premature by weight.	-	-	-	Sluggish.	Diff.to fix. Regurgitation.	Slow.	Severe right hemiplegia.
	170	Antepartum haem.	Foetal distress. Spont. vertex delivery.	6	-¾	Bronchopneumonia.	Mucousy.	Slapped. Aspirated.	2nd.day	3 weeks	Very restless and irritable.	Slow and difficult.	Poor.	Mild left hemiplegia. Bilateral optic atrophy. Mental defect.
	171	Rhesus incompatibility.	Occipito posterior. Spont. vertex delivery.	8	13	Normal.	Jaundiced.	Periodic	17th. day	-	Normal.	Normal.	Normal.	Severe right hemiplegia.

statements.

A Consideration of the Birth Histories of the 30 Patients With Congenital Hemiplegia.

From the descriptions which have been given of the abnormalities of pregnancy, labour and delivery encountered in the cases of congenital hemiplegia it is evident that these are rarely single. Abnormal pregnancy is more frequently followed by complicated labour and delivery than by normal labour and delivery, and commonly succeeded by an abnormal neonatal state. In 16 of the 23 patients born after abnormal labour or delivery, pregnancy had been abnormal, and in 19 of the 23 there was neonatal apnoea.

This point may be illustrated by considering some of the more frequently occurring abnormalities of parturition in more detail.

Pre-eclampsia was the commonest single abnormality of pregnancy. It was present in 10 cases. In only 3 of these patients were there no other complications of pregnancy and apparently normal spontaneous vertex deliveries. All 3 of these patients were apnoeic after birth. (Table 37). Pre-eclampsia was complicated by antepartum haemorrhage in two cases (Cases 100 and 201), both of whom were apnoeic after birth. In 4 cases labour was unduly prolonged (Cases 90, 111, 123 and 143), and in one there was a difficult low forceps delivery through a contracted pelvic outlet.

Similarly the majority of the 7 cases in which pregnancy was complicated by antepartum haemorrhage or threatened abortion had other disorders of pregnancy, labour or delivery. All /

TABLE 37

CONGENITAL HEMIPLEGIA

Case histories of patients whose mothers suffered from pre-eclampsia during pregnancy.

Case No.	Social Class	Mat. Age	No. of Preg.	Severity of PET	Other Complications	Mode of delivery	Placenta	Drugs	Bth.Wgt. lbs.	oz.	State of baby after birth.
43	IV	23	2	Moderately severe.	-	Spont. vertex	Unknown	-	7	-	Apnoea for some mins.
67	III	29	3	Severe.	-	Spont. vertex	Many infarcts.	Chloroform	8	1½	Apnoea for 20 mins.
176	III	29	1	Moderate.		Spont. vertex	Many infarcts.	Minnits Appar.	7	3½	Severe apnoea.
201	III	30	1	Moderate.	Antepartum haem. at term.	Spont. vertex	Many infarcts.	Minnits Appar.	6	2½	Apnoea for some mins.
90	IV	24	1	Severe.	-	Uterine inertia. Very prolonged labour. Failed forceps then successful high forceps.	Many infarcts.	Much pethedine. Ether. Spinal anaesthethia.	8	5	Apnoea for 15 mins.
100	IV	20	1	Moderate	Antepartum haem. immediately before labour.	Intrapartum haem. Spont. vertex	Clots and many infarcts.	Minnits Appar.	6	5	Apnoea for 10 mins.
111	V	29	1	Moderate	-	Uterine inertia. Foetal distress. Mid cavity forceps.	Many infarcts.	Cyclopropane. Pentothal.	7	14¾	Apnoea for 10 mins. Very cyanosed thereafter.
123	III	27	1	Severe	-	Surgical induction. Rigid soft parts. Prolonged labour. Spont. vertex deliv.	Normal.	Heroin and Chloroform.	7	-	Apnoea for some mins.
143	III	42	1	Severe	Backache.	Prolonged labour. Foetal distress. Mid cavity forceps for disproportion.	Offensive. Infected.	Morphia and Chloroform.	10	8½	Apnoea for some mins. Facial paresis.
86	I	28	1	Moderate	-	Low forceps delivery for outlet obstruction.	Unknown.	Chloroform.	7	4	2 teeth present but otherwise apparently normal.

TABLE 38

CONGENITAL HEMIPLEGIA

Case histories of patients whose mothers had revealed ante partum haemorrhage

Case No.	Social Class	Mat. Age	No. of Preg.	Time of haemorrhage	Other Complications	Mode of delivery	Placenta	Drugs	Bth. Wgt. lbs.	oz.	State of baby after birth.
5	V	40	3	Thirty five weeks.	-	Spont. vertex.	Unknown	None	4	12	Apnoea for some minutes.
70	V	45	4	Threatened abortion at 3 months.	-	Med. induction. Uterine inertia. High forceps deliv.	Unknown	Pituitrin. Morphia. Chloroform.	9	-	Regular respiration only after 2 hours.
74	V	26	1	Immed. before labour at 40 weeks.	-	Precipitate labour. Spont. vertex.	Unknown	None	6	3½	Apparently normal.
79	III	29	1	40 weeks. immediately before labour.	Hydramnios.	Cervical dystonia. Foetal distress. High forceps deliv.	Unknown	Pituitrin. General anaesthesia.	8	-¼	Artif. respiration for 1 hour.
100	IV	20	1	At 39 weeks.	Moderately severe pre-eclampsia.	Intrapartum haem. Spont. delivery.	Clots and many infarcts.	Minnits apparatus.	6	5	Apnoea for 10 mins.
109	IV	27	6	Threatened abortion at 4 and then 12 weeks.	Hyperemesis. Premature labour.	Spont. vertex.	Unknown.	None.	3	2	Very feeble. Apnoea for some minutes.
201	III	30	1	At 40 weeks. Day before delivery.	Pre-eclampsia.	Spont. vertex.	Many infarcts.	Minnits apparatus.	6	2½	Some minutes delay in breathing.

TABLE 39

Prolonged 2nd. stage of labour in cases with congenital hemiplegia

Case No.	Mat. Age	No. of Preg.	Pregnancy	Gest.	Delivery	Dur. of labour hrs.	Drugs	Condition of baby after birth	Bth.Wgt. lbs.	oz.	Placenta
162	42	6	Twin pregnancy. Severe anaemia. Cardiac failure	40	Prolonged 2nd.stage. Breech extraction.	24½	Chlorof.	Brief apnoea.	4	9	Normal
71	23	1	Normal.	40	Rigid soft parts. Foetal distress. Diff. high forceps.	48	General anaesth.	Apnoea 20 mins.	7	1¼	Unknown
180	21	1	Normal	40	Prolonged 2nd. stage. Low forceps deliv.	22	Chlorof.	Apnoea.	8	4	Infarcts
192	28	1	Normal	43	V. prolonged labour. Spont.vertex deliv.	120	Morphia Chlorof.	Apnoea.	8	12	Unknown
90	24	1	Mod. severe pre-eclampsia.	40	Uterine inertia. Forceps. deliv. at 2nd. attempt.	72½	Pethedine. Ether. Spinal anaesth.	Apnoea 15 mins.	8	5	Many infarcts.
123	27	1	Severe pre-eclampsia.	40	Surg. induct. Rigid soft parts. Prolonged labour.Spont.vertex.	37½	Heroin. Chlorof.	Prolonged apnoea.	7	-	Normal
143	42	1	Backache. Severe pre-eclampsia.	40	Prolonged labour. Foetal distress. Mid cavity forceps for disproportion.	48+	Morphia Chlorof.	Apnoea for only mins.	10	8½	Offensive
167	30	1	Recurrent pyelitis.	38	Prolonged labour. Spont. vertex deliv.	43½	Morphia	Apnoea for some mins. Premature.	5	4½	Unknown

TABLE 46.

Forceps delivery of children with congenital hemiplegia

Case No.	Mat. Age	No. of Preg.	Pregnancy	Gest.	Delivery	Dur. of Labour hrs.	Drugs	Condition of baby after birth	Bth.Wgt. lbs.	oz.	Placenta
71	23	1	Normal.	40	Rigid soft parts. Foetal distress. Diff. high forceps.	48+	General anaesthesia.	Apnoea for 20 mins.	7	1¼	Unknown
97	40	4	Normal.	40	Outlet contraction. Low forceps deliv.	9	Chlorof.	No apparent abnormality.	8	3	Unknown
180	21	1	Normal.	40	Prolonged uterine inertia. Low forceps delivery.	22	Chlorof.	"Some asphyxia"	8	4	Many infarcts.
70	45	4	Threatened abortion.	41	Med.induction. Uterine inertia. High forceps.	13	Pituitrin. Morphia. Chlorof.	Artif. Resp. 2 hrs.	9	-	Unknown
79	29	1	Hydramnios. APH at 40 wks.	40	Cervical dystonia. Foetal distress. High forceps deliv.	17½	Pituitrin. General anaesthesia.	Artif. Resp. 1 hr.	8	-	Unknown
90	24	1	Mod. severe pre-eclampsia.	40	Uterine inertia. Failed forceps. Succ. H. forceps at 2nd. stage.	72½	Pethedine. Ether.	Apnoea for 15 mins.	8	5	Many infarcts.
111	29	1	Mod. severe pre-eclampsia.	40	Uterine inertia. Foetal distress. Mid cavity forceps.	8¾	Cyclopropane. Pentothal.	Apnoea for 10 mins.	7	14¾	Many infarcts.
143	42	1	Backache. Severe pre-eclampsia.	40	Prolonged labour. Foetal distress. Mid cavity forceps for disproportion.	48	Morphia. Chlorof.	Apnoea for 30 mins.	10	8½	Offensive.
7	31	3	Fainting and nausea from 39 wks.	41	Dystonia. Mid cavity arrest. Mid cavity forceps. Diff. shoulder extraction.	2	Chlorof.	Normal.	9	13	Unknown.
86	28	1	Pre-eclampsia.	40	Low forceps for outlet arrest.	10	Chlorof.	Normal.	7	4	Unknown.

All but one of the patients was apnoeic after birth.(Table 38)

The second stage of labour was prolonged beyond 20 hours in 8 cases, the average duration being 52 hours. All the infants were first-born. All but one were apnoeic after delivery. Pre-eclampsia was present in 3 of the pregnancies. Congenitive cardiac failure, extreme post-maturity and recurrent pyelitis each complicated one pregnancy. Only two pregnancies (Cases 71 and 180) appeared to be normal. Prolonged labour was followed by assisted delivery in 5 cases, accompanied by foetal distress in two, and morphia or heroin was used in four (Table 39).

The birth histories of the 10 patients delivered by forceps are summarised in Table 40 . Only in 3 cases was pregnancy apparently uncomplicated (Cases 71, 97 and 180). Pre-eclampsia was present in 4 (Cases 90, 111, 143 and 86), antepartum haemorrhage or threatened abortion in 2 (Cases 70 and 79). It is of interest that patients showed signs of foetal distress prior to the forceps delivery in 4 cases (Cases 71, 79, 111 and 143) and had presumably suffered from some damage prior to the instrumental delivery. Six patients suffered from prolonged apnoea after delivery (Cases 71, 70, 79, 90, 111 and 143). To attribute the cerebral palsy to the effects of forceps delivery alone in these cases seems hardly justifiable, yet instrumental delivery has been regarded by many authors as a major cause of brain damage.

Twenty-four patients were apnoeic after delivery. Apnoea lasted for longer than 10 minutes in 12. Thirteen of the 24 patients were born after abnormal labour or delivery, and /

and in 17 pregnancies had been complicated. Thus neonatal apnoea may logically be regarded as a result of complications of pregnancy, labour and delivery in the majority of cases of congenital hemiplegia. Though it may often be an important contributory cause of foetal damage it cannot be considered the sole cause in any significant proportion of cases.

The impressive feature in the obstetric histories of patients with congenital hemiplegia are the constancy and multiplicity of the disorders of pregnancy, labour or delivery. Single abnormalities occur much less frequently than do a series of abnormalities. In the majority there are multiple potential causes of hypoxia and trauma. It seems to require severe multiple abnormalities of pregnancy labour and delivery to cause permanent damage in these patients.

Chapter 4c.

The Aetiological Findings in Patients with Acquired Hemiplegia.

Thirty three patients were classifed as suffering from hemiplegia acquired after birth. Twenty-two of them were male and eleven female, giving a similar distribution by sex to that found in congenital hemiplegia. The same hereditary, familial and social background was also apparent, except that birth rank tended to be lower and families larger than in congenital hemiplegia.

The ages at which the patients developed acquired hemiplegia are shown in Table 4I. It will be seen that approximately

TABLE 41

The Precipitating causes of acquired hemiplegia in 33 cases

Cause	AGE					Total
	0 - 6 mths.	6 - 12 mths.	1 - 3 yrs.	3 - 5 yrs.	5 + yrs.	
Cerebral thrombo-phlebitis.	0	0	1	0	0	1
Otitis media.	3	2	1	0	0	6
Pneumonia.	3	0	0	0	0	6
Dehydration; boils; osteomyelitis; dysentery.	3	0	0	0	1	4
Trauma.	1	1	0	2	1	4
Whooping cough (pertussis immunisation).	0	1	1	1	0	3
Measles.	0	0	1	0	1	2
Diphtheria.	0	0	1	0	0	1
Acute onset without apparent cause.	1	1	3	0	1	6
Slow insidious onset without apparent cause.	0	1	1	0	0	2
TOTAL	11	6	10	3	3	33

approximately half of the cases occurred in the first year, and in all but six cases was present before the age of 3 years. These figures are in accordance with the age distribution reported in most large series (Wyllie, 1952). It was estimated by Gowers (1888) that seven-eights of acute infantile hemiplegias occurred before the age of three years. Acute hemiplegia has generally been observed to be rare over the age of 7 years, and in fact only one case over this age was found in the present series (Freud, 1897; Gowers, 1888).

The birth histories of patients with acquired hemiplegia.

As has been stated, the criterion for the diagnosis of acquired hemiplegia was that there should be a history indicating that a child without signs of paralysis after birth developed signs of hemiplegia in post-natal life. Usually the hemiplegia resulted from an acute and recognisable illness, but in some cases the onset was insidious. In other cases the illness complicated by paralysis occurred within the first month of life and it was difficult to be quite certain that the child showed no motor abnormality at this early age. When there was any reasonable doubt about this, however, cases were classified as being of unknown origin. It is felt, therefore, that errors of classification in the series are probably few.

Nevertheless it is interesting to note the numbers of cases with a history of abnormal pregnancy, delivery and neonatal course, classified similarly to those with congenital hemiplegia (Table 42). Of the 33 cases of acquired hemiplegia /

TABLE 42

Abnormal pregnancy, labour, delivery and neonatal period in patients with acquired hemiplegia

Case No.	Place in family.	Pregnancy	Labour and delivery	State of child	Bth. Wgt. lbs.	oz.
89	3/5	Severe pre-eclamptic toxaemia.	Med. induction with pituitrin at 8 mths. Mid cavity forceps after 3 days in labour.	Apparently normal. Except for prematurity.	4	8
184	3/3	Severe bronchitis.	Normal labour of 7½ hrs. Spont. vertex delivery.	Normal	8	½
50	4/5	Mild pre-eclamptic toxaemia.	Spont. vertex delivery at term after 3 hrs. labour.	Normal	6	14
52	1/1	Mild pre-eclamptic toxaemia.	Spont. vertex delivery after labour of 13½ hours.	Normal	7	5½
165	1/3	Normal.	Assisted breech deliv. at 36 wks. gestation after labour of 22 hours.	Apnoea for few minutes.	Unknown	
20	5/6	Normal.	Prolonged labour of 48 hours at term. Spont. vertex deliv.	Normal.	8	3
164	2/2	Thyrotoxicosis. Hyperemesis. Bronchitis. Severe pre-eclampsia.	Delivered at 37 wks. spont. by the vertex.	Normal.	5	12
188	8/9	Ill and tired. Haemoglobin 50%. Hydramnios.	Spont. vertex delivery at 30 weeks after 12 hours.	Cyanotic episode after breathing normally after birth.	3	7

hemiplegia parturition was abnormal in some respect in 9. In 4 patients pregnancy only was abnormal (Cases 184, 50, 52 and 164). In 3 labour or delivery or both were abnormal though pregnancy was normal (Cases 24, 165 and 20). Two infants were premature by birth weight (Cases 89 and 188), the latter having cyanotic episodes after birth, and two were apnoeic for a few minutes after birth. Only one case showed abnormalities of pregnancy, delivery and the neonatal period (Case 89).

When the cases are analysed in a similar fashion to those with congenital hemiplegia it is found that 5 had evidence of possible hypoxia during pregnancy or during labour and delivery, and 3 had evidence of possible or probable trauma. Only 2 patients had evidence of possible or probable trauma with possible or probable hypoxia (Table 42). The marked differences in the birth histories of these patients and of those with congenital hemiplegia are evident. In view of this and the fact that all the patients were apparently normal for some time after birth, it seems unlikely that disorders of parturition were the cause of hemiplegia in the group of patients labelled acquired. On the other hand the possibility of there being congenital cerebral abnormalities as a result of hereditary factors or disorders of parturition which made the child's brain more liable to be damaged by trauma or disease occurring in later childhood cannot be ruled out. This possibility has particularly to be remembered in view of the finding that congenital anomalies and a positive family history of mental and neurological disease were found equally /

equally frequently in patients with congenital and acquired hemiplegia.

Types of onset.

In 26 cases acquired hemiplegia occurred as a complication of a previously diagnosed illness, or as a result of trauma. The illnesses will be discussed in more detail later and are indicated in Table 43 . In 7 cases the hemiplegia occurred without any diagnosis of pre-existing illness being made (Table 44).

Two modes of onset could be distinguished in the cases of acquired hemiplegia, whether or not the condition complicated a prior illness. The majority of the patients, 28 of the 33, suddenly showed signs of acute disorder of the nervous system, most frequently convulsions with loss of consciousness, followed by manifest hemiplegia as soon as consciousness was restored. A few patients with such an acute onset did not have convulsions but were very restless, irritable or drowsy for a period of hours before the hemiparesis became apparent. In 5 patients the hemiplegia was less dramatic in its appearance, and in these cases the term sub-acute is used to describe its onset. The condition became apparent in these over a period of hours or days after days or weeks of illness.

The modes of onset of acute and sub-acute acquired hemiplegia will not be considered in more detail.

Acute acquired hemiplegia.

In 4 cases acute hemiplegia followed cranial trauma, in 19 it occurred as a complication of previously diagnosed disease, /

TABLE 44.

The history of acquired hemiplegia in patients without identified pre-existing illness

A. ACUTE ONSET

Case No.	Social Class	Place in family	Age at onset of neurological disorders.	Previous health	Neurological disorders.	When hemiplegia noted
20	V	5/6	2 yrs. 7 mths.	Good.	Sudden generalised convulsion lasting 12 hours.	After convulsion.
85	III	1/2	5 years.	Good.	Complained of sudden 'dizziness' and fell aged 5. Thereafter headaches after one month.	After fall.
94	V	2/2	2 yrs. 11 mths.	Good.	Generalised convulsions aged 2 yrs. 11 mths. with loss of consciousness. Well thereafter. 6 wks. later very severe generalised convulsions with cyanosis. Unconscious 4 days.	After 2nd. convulsion
121	III	only	15 months.	Good apart from severe chronic tonsillitis.	Severe generalised convulsions for 2 hrs. with marked cyanosis and loss of consciousness. Convulsions every 3 mths. thereafter.	After generalised convulsion at 30 mths.
142	III	2/4	7 months.	Well apart from irritability and fretfulness 5 days before convulsion.	Convulsion confined to right side with loss of consciousness. Lasted 10 mins.	After convulsion.
B. SUBACUTE ONSET						
11	III	1/3	12-15 mths.	Well.	Gradually began to use left arm and leg less and less between 12 and 15 months.	Between 12 and 15 months.
175	V	1/3	9 months.	Mild fever of unknown origin for some weeks.	Began to drag left leg when crawling and then ceased after some days to use left arm.	About $9\frac{1}{2}$ months.

TABLE 45.

Acute acquired hemiplegia with convulsions at onset

Case No.	Age in Yrs.	Mths.	Prior illness or neurological signs and symptoms.	Day of convulsion	Type of initial convulsion	Frequency of subsequent attacks	When hemiplegia apparent.	Epilepsy later
17	6	52	Pneumonia and mastoiditis. Restlessness. Neck stiffness.	3	Shocked with loss of consciousness for few moments.	Many times a day. Similar to first attack.	Within day or two of attack.	+
24	-	9	Bilateral otitis media.	10	Unilateral with loss of consciousness for some minutes.	No subsequent attacks.	After fit.	-
93	-	9	Otitis media.	1	Unilateral with loss of consciousness for half an hour.	No subsequent attacks for 1 year.	After fit.	+
144	1	10	Otitis media.	7	Generalised with loss of consciousness for many hours.	Once every 2-3 days for few months.	After initial fit.	+
50	-	3	Otitis media. Meningitis.	14	Generalised without loss of consciousness lasting 5-10 minutes.	6-12 times in 24 hours up to 7 mins. each. Similar to first attack for 3 mths.	After a number of convulsions. Several days after onset.	+
188	2	8	Recurrent pneumonia.	1	Unilateral with loss of consciousness for some hours.	"One or two some days".	After first fit.	+
6	-	3	Pneumonia.	7	Generalised but more on left. Loss of consciousness for 2 hours.	Up to 20 times in day for up to 20 mins. for 2 days.	After first fit.	-
164	-	5½	Recurrent pneumonia.	2	Generalised but more on left. Loss of consciousness intermittent for 36 hours.	Intermittent 10-15 times a day for some weeks. Momentary.	After 2 - 3 days.	+
19	1	3	Whooping cough.	Few days	Unilateral with loss of consciousness for 10 minutes.	Similar to 1st. attack. 3-4 times a day.	After 1st. fit but none after later attack.	+
69	3	-	Whooping cough.	7	Generalised with loss of consciousness for few moments.	18 in next 24 hrs. and then 4-5 a day like first attack.	After 3 weeks of fits.	+
196	2	6	Whooping cough.	28	Generalised with loss of consciousness intermittently for 24 hours.	6-7 times a day for 4 days for 3-4 months. Generalised.	After first fit.	+
191	5	-	Measles.	6	Generalised with loss of consciousness for a few minutes.	No subsequent attacks.	After convulsion.	-
184	2	6	Measles and pneumonia.	Few days	Generalised with loss of consciousness for few moments.	Three similar in next three days.	After 1st. attack but none after 3rd.	-
85	5	-	Intermittent headaches.	30 days	Unilateral. Impairment of consciousness for few minutes.	No subsequent attacks.	After convulsion.	-
142	0	7	Fretful and irritable. Impairment of vision.	5	Unilateral with loss of consciousness for 10 mins.	No subsequent attacks.	After convulsion.	-
94	2	11	-	1	Generalised with loss of consciousness for 2 hrs.	Similar 6 wks. later. Thereafter one every 3 months.	After second convulsion.	+
121	1	3	-	1	Generalised with loss of consciousness for 3 hours.	Similar attacks every 3 mths. sometimes more marked on Rt.	After initial convulsion.	+
20	2	7	-	1	Unilateral with loss of intermitt. consciousness 12 hrs.	No subsequent attacks.	After initial convulsion.	+
48	18½ wks.		-	1	Generalised with loss of consc. unknown period & then for 48 hrs.	Gen. attacks with loss of consc. 2-3 times a day for few minutes.	After initial convulsion.	+

disease, and in 5 it appeared in children who were apparently healthy. The cases resulting from trauma will be considered later after the other 24 cases have been discussed (Tables 45 - 46).

Cases with Convulsions. In 16 of the 24 cases convulsions were the first sign of acute neurological involvement. In 8 cases convulsions were not the first sign of neurological involvement, and 5 of these did not have fits at the time of the onset of their hemiplegia. Three cases had convulsions some days after other signs of nervous disorder had been present. In Case 191 drowsiness and head retraction were present for one week, in Case 85 intermittent headaches for 30 days, in Case 142 fretfulness and impairment of vision for 5 days. In the 5 cases without convulsions at the time of the onset of the hemiplegia, the hemiparesis was the first sign in three cases (Cases 89, 110 and 132), neck stiffness and drowsiness in Case 129, and restlessness and impairment of consciousness in Case 169. (Table 45)

Thus 19 cases had convulsions at the time of the onset of their hemiplegia, 16 as the first sign of nervous disorder and 3 after other signs had presented. The form of the convulsion was generalised twitching of the limbs with loss of consciousness in 12 cases, in 2 of which the limb movements were more marked on the side later paretic than on the other (Cases 6 and 164). In all these cases the clonic phase of the fit was much more marked than was the tonic. In some patients who were admitted to hospital and whose fits were described, the tonic phase is not even mentioned. More frequently /

frequently recurrent clonic attacks were interspersed by periods of flaccidity and persistent impairment or loss of consciousness. One patient (Case 50) had generalised involuntary clonic movements of the limbs with severe impairment but no loss of consciousness. In 6 patients movements were reported to be confined to the side which later showed paresis. In 5 of these there was loss of consciousness, and in one, impairment.

The duration of the fits was extremely variable. They were momentary in some patients and lasted as long as 24 hours in others (Cases 196 and 48 for example). Convulsions were probably single in 6 cases and recurrent in 13, though it is difficult in some to be sure whether persistent loss of consciousness was due to heavy anti-convulsive sedation or to persistent epileptic phenomena.

Following the initial fit the hemiplegia was almost invariably noted to be present for the first time and usually the affected limbs were stated to have been found flaccid and useless, "dead" and often described as being discoloured and swollen, and hot or cold. In all the patients with single convulsions the hemiplegia was most severe in the period immediately following the convulsion and thereafter a gradual improvement was noted. In children with more than one fit, however, the hemiplegia might apparently be more severe after a subsequent episode. In Case 94, for example, a boy aged three showed a typical mild left hemiplegia following a generalised convulsion lasting 2 hours without a history of preceding illness. The hemiplegia gradually improved during the /

the next six weeks so that some use of the affected hand was possible, but following another similar convulsion lasting three hours at this time became much more severe. The limited use of the hand apparent before the second convulsion was never recovered.

In summary, the most frequent course of events in these patients was a unilateral or generalised convulsion with loss or severe impairment of consciousness either in the course of a pre-existing illness or when he is apparently healthy. The convulsion might last only a few minutes or for as long as 24 hours. It might be single or be followed by other convulsions. The hemiplegia was usually apparent immediately after the first convulsion and the paresis was most marked then but occasionally became more severe after subsequent attacks.

Acute Onset Without Convulsions. Five cases developed hemiplegia without having convulsions at the time of onset. In all the paresis complicated the diseases. Case 10 was noted to be normal immediately after birth, but became desperately ill at the age of 3 days with mastoiditis. The paresis was noted one day between on and two weeks after the onset of the illness. Case 89 was very severely ill with acute osteomyelitis of the tibia, and in the course of 24 hours became paretic on the right side and lost the power of speech about the 10th day of his illness. (Table 46)

Case 129 was ill for a period of 4 months with palatal paralysis complicating his diphtheria. For a week during the earlier part of his illness he was noted to have neck stiffness and drowsiness, and shortly afterwards a left hemi-

paresis /

TABLE 46.

Acute onset of acquired hemiplegia without convulsions

Case No.	Age at onset of illness	Nature of illness	First signs of neurological disorder	Time in illness when diplegia noted.
110	3 days	Mastoiditis.	Poverty of right sided movement. Restlessness. Irritability.	Between 1 and 2 weeks.
89	8 years.	Acute osteomyelitis.	Right hemiparesis.	About 10 days.
129	1 yr. 9 mths.	Diptheria.	Neck stiffness. Drowsiness. Left hemiparesis.	Within 2-4 weeks.
132	3 months.	Dysentery.	Left hemiparesis.	2 days.
169	1 yr. 1 mth.	Measles. Pneumonoccal septicaemia.	Restless and loss of consciousness followed by paresis within 3 weeks.	2nd. day of illness.

hemiparesis was apparent. Case 132 was admitted to hospital with dysentery at the age of 3 months and suspected of having meningitis in addition. A left hemiparesis was noted at the time of admission on the second day of the illness. Case 169 developed measles which was complicated by pneumococcal meningitis. The initial signs of neurological involvement were impairment of consciousness and restlessness. He was admitted to hospital on the second day of his illness and found to show a slight right hemiparesis which became worse in the course of the following two or three days.

It is of interest that though none of the 5 patients had convulsions at the time of the development of his hemiplegia, two (Cases 110 and 169) subsequently developed epilepsy. In Case 110 the first fit occurred shortly after the age of one year, and in Case 169 at the age of 16 months, 4 months after the onset of his illness.

Acute Onset With Trauma. Four cases, all, oddly enough, female, showed hemiplegia following trauma (Table 47). Two were under one year of age and the others aged 3 and 4 years (Cases 195 and 52 respectively).

All 4 cases suffered falls onto the head. Case 4 was dropped on the head at the age of 4 months while her parents were escaping from Poland in the early days of the war. Whereas the child had previously been alert and lively, feeding well and showing keen interest in her surroundings, within a few hours of her accident she seemed drowsy and next day was drowsy, fretful and refused to feed, and had difficulty in swallowing. Within a few days it was apparent that /

TABLE 47.

Histories of patients with acquired hemiplegia after trauma

Case No.	Age	History	First signs of neurological disorder	Appearance of hemiplegia
15	10 mths.	Fell from pram on to head.	Twisted face. Hemiparesis.	At once.
52	4 yrs.	Fall and penetrated left carotid artery.	Generalised fit following fall, followed by hemiplegia.	At once after fit.
165	4 mths.	Fall from cart on to head.	Drowsiness and refusal to feed next day.	Within one week.
195	3 yrs.	Fall from air raid shelter.	Loss of consciousness for 2 days.	Within 2 days on recovering consciousnes

that the right limbs were moved less than the left. Epilepsy developed at the age of 4 years, but no fits were noted prior to this time.

Case 15 fell from her pram at the age of 10 months when a little boy tried to move her pram. A few minutes later she was picked up and noted to be "dopey", not apparently recognising her parents and was found to have a twisted face. The left limbs were observed to be moved less than the right later the same day. She was admitted to hospital and discharged after one week diagnosed as suffering from the effects of concussion.

Case 195 fell on her head on to concrete from an air-raid shelter at the age of 3. She was picked up unconscious and admitted to hospital, where concussion and an extensive fracture of the skull were diagnosed. She was unconscious for 2 or 3 days and on recovery a left hemiplegia was found. One year after her accident epileptic attacks commenced.

Case 52, whose mother was epileptic, fell from a sofa with a pencil in her mouth at the age of 4. She immediately had a generalised convulsion and was admitted to hospital unconscious and in a state of generalised hypertonicity. A penetrating injury of the left tonsillar bed was found. She recovered consciousness after 1½ hours but could not speak, and a flaccid right hemiparesis was observed. Four months after her injury there was considerable improvement in movements of the right limbs. Aphasia was still apparent. She had had one generalised epileptic fit and subsequently other similar fits occurred.

Subacute /

TABLE 48.

Subacute onset of acquired hemiplegia

Case No.	Place in family	Social Class	Age at onset	Nature of preceding illness	Duration of illness	Fits and type	When hemiplegia noted
4	1/1	II	1/0	Diphtheria and pertussis immunisation followed by altered state of consciousness. Fever and whimpering.	8 weeks	None.	Aged 14 mths. Ceased to use first right arm and then leg.
11	1/3	III	1/0 - 1/3	None.	-	None.	Gradually ceased to use left limbs between 12 and 15 months of age.
117	1/1	II	/2 wks.	Pyloric stenosis. Very severe dehydration.	5 weeks.	Generalised from aged 3 weeks.	Aged 3 months. 3 weeks after discharge from hospital.
175	1/3	V	/9	Pyrexia of unknown origin aged 9 months.	weeks.	None.	Began to drag left leg when crawling and then ceased to use left arm aged 9 months +.
187	1/7	III	/1	Boils.	Several weeks.	None.	Gradually ceased to use right limbs.

Subacute Onset of Acquired Hemiplegia.

In five patients the onset of hemiplegia was subacute rather than acute in type. (Table 48.) In two patients (Cases 11 and 175) there was no apparent cause of the hemiplegia. Case 11 gradually used the left arm less and less and began to drag the left leg, over a period of about three months from the age of one year. The child was not apparently ill during this period and after the age of 15 months there was no suggestion that the paralysis was progressive in type. Case 175 was fretful and anorexic and slept poorly for a period of 3 or 4 weeks at the age of about nine months, and towards the end of this time it was noted that the child dragged the left leg when crawling and was unable to use the arm as strongly as the right.

Case 117 became very dehydrated by the age of three weeks as a result of pyloric stenosis and brief lapses of consciousness were noted 5 - 6 times a day from this time until the child was 5 weeks old, when she was admitted to hospital for operation. On admission the right limbs were noted to be moved much less freely than the left.

Case 4 developed an acute cellulitis following the injection of mixed pertussis and diphtheria vaccine and was fevered and restless for approximately 2 months, at the end of which time the right arm was noted to be flaccid and useless though the leg seemed unaffected. One week later the leg also became flaccid and useless in a rather similar manner to the arm.

In Case 187 the mother developed septic mastitis following discharge /

discharge from maternity hospital, and the infant had a series of boils from the age of 2 - 6 weeks. During this time he was fretful and fevered and a right hemiplegia was noted some time between the 4th and 5th weeks, which waned gradually over a period of a few days.

It is of interest that only one patient had epileptic manifestations during the period of the subacute onset of hemiplegia (Case 117) and that this patient was the only one to have subsequent epileptic attacks.

Conditions Complicated by Hemiplegia.

Otitis Media. In Table 49 are shown the clinical summaries of 6 patients who developed hemiplegia whilst suffering from otitis media or mastoiditis. All but one, Case 110, had convulsions. In Case 17 the mastoiditis was associated with increased intracranial pressure, and in Case 50 it was followed by meningitis. Unfortunately it was not always possible to determine the date of the onset of the otitis in these cases accurately. The stated day after the onset of illness, on which the first indications of neurological disorder were evident, is therefore not reliable, but probably gives some indication of the lapse of time between symptoms and signs of ear disease and signs of nervous disturbance being apparent. It will be observed that in only one case was this period less than 5 days and in no patient did it exceed 14.

The otitis media was bilateral in Case 50 and unilateral in the other five. In 2 cases (Cases 110 and 17) there was mastoiditis, and this was treated by penicillin injections in the /

TABLE 49.

Otitis media complicated by hemiplegia

Case No.	Age at onset	Remarks	Treatment	Complications	Neurological disorders
50	3 months.	Bilateral.	Nil.	Streptococcal meningitis aged 3½ months.	Convulsions at 3½ months and right hemiplegia.
93	9 months.	Severe right otitis media which had been present for some time.	Nil.	-	Convulsions at 9 months, left hemiplegia.
110	3 days.	Acute left mastoiditis.	Penicillin.	Severe dehydration.	Right hemiplegia after four days.
144	4 months.	Chronic right otitis media.	Local treatment.	Acute upper respiratory infection and acute otitis media on right. Aged 22 mths. Right jugular thrombosis.	Convulsions aged 22 mths. Left hemiplegia.
17	5 weeks.	Mother developed staph. aureus abscess at 4 wks. and 10 days later infant dev. staph. otitis media and mastoiditis on left.	Left Schwartz operation.	Staphylococcal cerebral thrombophlebitis and meningitis at 7 weeks.	Convulsions aged 5 mths. and right hemiplegia.
24	9 months.	Streptococcal otitis media on unknown side.	Sulphonamides.	-	Convulsion 10 days after onset and left hemiplegia.

the former and a Schwertz operation in the latter. In Case 24 sulphonamides were given to the patient with streptococcal otitis media. There was severe dehydration in Case 110 which was corrected by saline intravenous and subcutaneous infusions. In Case 144 local treatment of chronic right otitis media appeared to have little effect, and when the child acquired a cold at the age of 22 months acute otitis media resulted. This case was of special interest in that a jugular thrombosis on the right could be demonstrated clinically. Involvement of the cerebral veins was demonstrated on exploratory craniotomy in Case 17. Case 17 and Case 50 suffered from complicating meningitis, staphylococcal in the former and streptococcal in the latter. Unfortunately cerebro-spinal fluid findings are not available for all the cases. Cases 93 and 110 did not have lumbar puncture. In Cases 144 and 24 lumbar puncture findings were normal, and in Cases 50 and 17 high polymorph counts with organisms in the fluid were reported.

Pneumonia as a Cause of Acquired Hemiplegia. Three cases had pneumonia during which generalised convulsions occurred and were followed by the appearance of hemiplegia. (Table 50) All the patients were under the age of 6 months. Case 188 was born prematurely and had repeated attacks of pneumonia from the age of 10 days until the time of examination at the age of 4 years. During his second attack he had a number of extremely severe convulsions accompanied by very marked cyanosis. He was admitted to hospital apparently dead. Severe bilateral bronchitis was diagnosed and treated with streptomycin /

TABLE 50.

Pneumonia complicated by acquired hemiplegia

Case No.	Age at onset of illness	Illness	First signs of neuological involvement.	Paralysis
6	3 months.	Cold followed by pneumonia after one week.	Convulsions during first two days of pneumonia. Generalised but most marked on left side.	Left hemiplegia.
164	5½ months.	Chronic bronchitis following whooping cough aged 9 weeks. Bronchopneumonia aged 5½ months.	Generalised convulsions on 2nd. day of bronchopneumonia.	Left hemiparesis.
188	21 weeks	Pneumonia aged 10 days and again aged 21 weeks.	Generalised convulsions on first day of illness.	Right hemiplegia.

streptomycin, aureomycin and penicillin. After he regained consciousness it was noted that the right limbs were paretic.

Case 164 developed whooping cough aged 9 weeks, and thereafter had a chronic cough and nasal catarrh. At the age of 5½ months the catarrh and cough became more marked and bronchopneumonia was diagnosed. The child was treated with penicillin, but the following day had a severe generalised convulsion lasting 36 hours, and was admitted to hospital with a left hemiplegia. Lumbar puncture showed a fluid pressure of 280 mm. C.S.F. but constituents were normal. The child recovered after treatment with chloromycetin and oxygen. Case 6 was diagnosed as suffering from lobar pneumonia after having had a cold for about one week before becoming very ill. During the first two days of her illness she had a number of generalised convulsions with very severe cyanosis which lasted as long as two hours. Following one particularly severe attack which marked the left limbs more than the right she was noted to show a left hemiplegia. She recovered after treatment with penicillin injections.

Meningitis. Hemiplegia developed in Case 169 at the age of 13 months. The child developed measles, and 2 weeks after the rash appeared became drowsy and lethargic, fevered and restless at times, and was admitted to hospital where a mild right hemiplegia was found which became more severe in the course of the next few days. Lumbar puncture was performed and a pure culture of pneumococci obtained from the fluid. Later an intrathecal block developed. The child became deaf, partially blind, aphasic, and suffered from recurrent epileptic /

epileptic sequences in addition to his right hemiplegia. He was one of the first civilian cases in Edinburgh to be treated with penicillin.

Whooping Cough. Three patients (Cases 69, 19 and 196) had whooping cough complicated by hemiplegia; they were aged 3 years, 15 months and 2½ years respectively. (Table 51) Case 69 had relatively mild symptoms of whooping cough but there was evidence of chronic tonsillar infection. Eighteen generalised convulsions occurred in 24 hours, after cough had been present for one week. These were suppressed by phenobarbitone, but recurred 4 - 5 times a day after 3 weeks' freedom. During the second series of fits left hemiparesis became apparent. Papilloedema was present. Lumbar puncture findings were normal. He was treated with penicillin for a period of months, but considerable mental impairment ensued and occasional fits continued to occur.

Case 19 was admitted to hospital with a unilateral convulsion with loss of consciousness after being vaguely off colour for a few days. On recovering consciousness she was found to have a left hemiplegia, and whooping cough was diagnosed. Lumbar puncture findings were normal.

Case 196 had very severe whooping cough with such severe bouts of coughing that she would lie on the floor gasping for breath and blue in the face during them. After 4 weeks she developed screaming attacks and appeared hallucinated during them. After 3 days generalised convulsions ensued and she was admitted to hospital where a left hemiplegia was noted. Lumbar puncture findings were normal.

Measles. /

TABLE 51.

Whooping cough and measles complicated by acquired hemiplegia

Case No.	Age at onset	Clinical details	Neurological disorders	Paralysis
MEASLES				
184	14 months.	Admitted to hospital on account of bronchopneumonia complicating measles and developed encephalitis with many convulsions.	Drowsiness and convulsions. Lumbar puncture. Findings unknown.	Right hemiplegia after first convulsion.
191	5 years.	Became very drowsy, developed neck stiffness 5 days after the appearance of measles rash.	Drowsiness, disorientation. Convulsions 2 wks. after appearance of rash. Normal lumbar puncture findings.	Right hemiplegia after first convulsion.
WHOOPING COUGH				
69	3 years.	Relatively mild whooping cough. Enlarged tonsils and cervical glands.	Generalised convulsions after 1 week and 18 subsequent fits in next 24 hours. Then 3 wks. freedom followed by 4-5 fits every day. Lumbar puncture findings normal.	Left hemiparesis developed during second series of fits.
19	15 months.	Admitted to hospital with convulsions having been fevered and off colour for a few days. Severe whooping cough diagnosed.	Unilateral convulsions 3-4 times a day.	Left hemiplegia apparent after first convulsion.
196	2½ years.	Very severe whooping cough. Seemed more ill after 4 weeks and had screaming attacks for some days and then convulsions. Admitted to hospital.	Restlessness and irritability and generalised convulsions. Lumbar puncture findings normal.	Left hemiplegia apparent after first convulsion.

Measles. Two patients had measles (Cases 184 and 191), aged 14 months and 5 years respectively. Case 184 was admitted to hospital on account of complicating broncho-pneumonia and drowsiness. Measles encephalitis was diagnosed. Whilst in hospital he had many generalised convulsions, and after the first of these right hemiplegia was noted. Lumbar puncture findings were unknown.

Case 191 became very drowsy and disoriented and developed neck stiffness 5 days after his measles rash had appeared. He was admitted to hospital with a diagnosis of measles encephalitis. Lumbar puncture findings were normal. After one week in hospital he had a generalised convulsion, and after it a right hemiplegia was noted. He was unconscious for one week after the fit, but thereafter steadily improved. (Table 51)

Diphtheria. One child (Case 129) developed hemiplegia whilst suffering from severe diphtheria with palatal paresis and endocarditis. Three weeks after the onset of the infection he became delirious and developed head retraction which lasted for one week. During this time lumbar puncture findings were normal, but a left hemiplegia became apparent.

Dysentery. One child (Case 132) had very severe dysentery and was admitted to hospital very dehydrated after 24 hours. He was treated by subcutaneous and intravenous infusion, and when somewhat recovered was found to have a flaccid left hemiparesis not present on previous hospital admission 3 weeks previously. Primary pulmonary tuberculosis was also present.

Acute /

Acute Osteomyelitis. Acute osteomyelitis was present in one child (Case 89) who developed right hemiplegia after 10 days' illness, during which time he had been operated upon in order to draw pus from the lower end of his right tibia. The boy was severely ill in hospital and was treated with penicillin for more than three months.

Vaccination. The boy in Case 48 was found unconscious, face down in the pillow of his pram 17 days after vaccination, and 8 hours after he had apparently been successfully resuscitated went into status epilepticus for 24 hours. After this a severe right hemiplegia was noted.

Cases Without Apparent Cause. The case histories of the 5 acute and 2 subacute cases of acquired hemiplegia in which the cause of the disorder was obscure are covered in Table 44 . In all but one the milestones were apparently normal. In Case 94 walking without support was not achieved until 15 months and talking until 20 months. Only two patients, one (Case 20) when the onset was acute, and one (Case 111) when it was subacute, showed no evidence of predisposing illness. Case 20 had a sudden generalised convulsion at the age of $2\frac{1}{2}$ years with loss of consciousness for 12 hours intermittently, after which the hemiplegia was apparent. Case 111 gradually ceased to use the left limbs during a period of about 3 months at the age of 12 to 18 months.

Three cases (Cases 121, 94 and 85) had convulsions some weeks before developing hemiplegia. In Case 121 generalised convulsions lasting as long as 2 or 3 hours with loss of consciousness occurred every three months from the age of 15 months /

months at a time when the child had very large inflamed tonsils and cervical adenitis. At the age of 30 months a rather more severe convulsion was followed by the appearance of a right hemiplegia and severe mental impairment. Case 94 had a severe generalised convulsion with loss of consciousness for 2 hours at the age of 2 years and 11 months in association with sudden diarrhoea and vomiting. Six weeks later another severe generalised convulsion lasting 4 hours occurred after he had seemed off colour for a few hours, and on recovery a left hemiplegia was apparent. Case 85 felt dizzy and fell at the age of 5. For four weeks after he complained of severe intermittent headaches and then had a convulsion confined to the left side, with impairment but no loss of consciousness. On admission to hospital a left hemiplegia was found.

Case 142 was fevered, anorexic, irritable and lethargic for 5 days prior to having a right sided convulsion with loss of consciousness at the age of 10 months. Bilateral papilloedema and right hemiplegia were noted on recovery.

Case 175 seemed fevered and fretful for some weeks at the age of 9 months and ceased to gain weight. He began to drag his left leg when crawling and after some days ceased to use the left arm, and was found to have a left hemiplegia on examination.

Probable Clinical Pathology.

Intracranial Thrombosis or Thrombophlebitis. The retrospective diagnosis of probable intracranial thrombosis or thrombophlebitis was based on circumstantial evidence in the majority /

TABLE 52.

Probable intracranial thrombophlebitis as a cause of acquired hemiplegia

Case No.	Age of onset of illness	Preceding illness	Onset of neurological abnormalities	Course of illness and treatment
17	4 weeks.	Pneumonia and mastoiditis. (Mother had septic mastitis) with dehydration and multiple abscesses.	Fifth day. Refusal to feed. Stiff neck.	Rt. mastoidectomy age 6 wks. Cerebral abscess drained age 8 mths. after recurrent convulsions in the interim.
24	9 months.	Bilateral otitis media.	Tenth day. Left sided convulsions with loss of consciousness.	Otitis was treated with penicillin in Fever Hospital.
93	9 months.	Otitis media.	First day. Fit followed by left hemiparesis.	Treated with penicillin injections as out-patient.
110	3 days.	Mastoiditis.	7-8 days right hemiplegia. Retarded milestones thereafter.	Treated with sulphonamides and penicillin. Lost 1 lb. in weight during illness.
144	22 months.	Otitis media.	At 7 days had a convulsion followed by left hemiplegia.	Developed palpable right jugular thrombosis whilst in hospital on treatment with penicillin. Persistent convulsions.
50	3 months.	Otitis media.	14 days after otitis noted. Meningitis with convulsions.	Haemolytic streptococcal meningitis found on admission to hospital. Treated with penicillin but residual hemiplegia.
169	13 months.	Measles. Pneumococcal septicaemia and meningitis.	Restlessness and severe coma 2 weeks after developing measles and 2 days after massive cervical adenopathy.	Found to be suffering from pneumococcal meningitis 6 days after adenopathy noted. Penicillin systemically and intrathecally.
89	8 years.	Acute osteomyelitis. Pericarditis and pneumonia.	Right hemiparesis within 2 weeks.	Osteomyelitis treated by surgical drainage. Hemiplegia noted after a few weeks.
187	2 months.	Recurrent multiple boils. (Mother had breast abscesses).	Fretful and showed right hemiparesis after about 2 weeks.	Local treatment to boils only. No systemic treatment.
85	5 years.	Fell and complained of headache and dizziness.	Intermittently for one month.	Hemiplegia appeared after he had been ill for one month.
142	7 months.	Seemed out of sorts and fevered for 5 days. Bronchitis for 4 weeks beginning 6 weeks before.	Convulsion confined to right side for 10 minutes.	Right hemiplegia noted after convulsion.
175	10 months.	Pyrexia of unknown origin for some weeks.	Began to drag left leg and then arm some weeks after onset of illness.	Pyrexia settled gradually after some weeks by which time some return of function in paretic left limbs.
117	2 weeks.	Pyloric stenosis with very severe dehydration.	Generalised convulsions aged 3 weeks.	Ramstedt's operation aged 5 weeks. Gradual improvement but hemiplegia noted age 3 months, 2 weeks after discharged from hospital.
111	12-15 mths.	None.	Gradually ceased to use left limbs between 12 and 15 months of age.	Gradually attained some return of function after 15 months.
132	3 months.	"Dysentery" with severe dehydration.	Noted to be hemiplegic when temperature settled. (No convulsions).	Treated with sulphonamides and I.V. fluids successfully but hemiplegia persisted.
4	12 months.	Third diphtheria and pertussis immunisation 8 weeks before had been followed by persistent listlessness, anorexia and fever.	Paresis first evident in right hand. Spread to involve lower limb within 3 days.	Severe hemiplegia was considerably improved by end of 4 weeks.

majority of cases. Even at the time of the acute illness it is not very easy to make a definitive diagnosis and it is even more difficult to make one on the basis of second-hand reports. Nevertheless sufficient evidence can be adduced from the histories and clinical courses of the patients in the majority of cases to justify at least a tentative diagnosis in 16 of the 33 patients suffering from acquired hemiplegia (Table 52)

In 10 cases there was a history of previously diagnosed pyogenic infection - otitis media or mastoiditis in 6 (Cases 17, 24, 93, 110, 144, 50) all but one of whom were under the age of 9 months, pneumococcal septicaemia and meningitis in Case 169, acute osteomyelitis, with pericarditis and pneumonia in Case 89, multiple carbuncles in Case 187, and dysentery with severe dehydration in Case 132.

In 3 further cases there was pyrexia of unknown origin (Cases 4, 42 and 175), and in 3 (Cases 85, 111 and 117) there was no history of preceding infection. Case 117 had hypertrophic pyloric stenosis and was marasmic and very severely dehydrated before efficient treatment was commenced.

It is interesting to compare this series of patients with acquired hemiplegia in childhood with the series of cases of sinus thrombosis reported by Byers and Hass (1933). Adopting their classification of cases, two (Cases 117 and 132) would be classified as "Primary thrombosis" (also called marantic thrombosis), since both were severely dehydrated babies (under the age of 30 months) at the time of the onset of acute neurological abnormalities. Nine would be considered to be cases of infective thrombosis, and, as in their series of /

of sinus thrombosis, the commonest preceding infection is middle ear disease or mastoiditis. In Case 187 multiple carbuncles due to staphylococcal aureus infection over a period of 2 months would be a history quite compatible with intracranial septic thrombosis. In Case 89 the gradual appearance of hemiparesis in the course of acute staphylococcal osteomyelitis, pneumonia and pericarditis argues in favour of venous thrombosis rather than septic embolism in which a more acute onset of hemiplegia is more frequent. But "toxic encephalitis" is another possibility here which cannot be ruled out in retrospect.

In Case 142 the girl aged 7 months had been well apart from a prolonged attack of bronchitis from which she had appeared to have recovered two weeks before the onset of her acute illness which resulted in the occurrence of hemiplegia. She was noted to be progressively drowsy, irritable and fevered for 5 days prior to her being admitted to hospital where she was found to be semi-conscious, fevered, and to have bilateral papilloedema and constant nystagmus to right, and a white blood count of 13,000. Shortly after admission she had a convulsion confined to the right side. Following it there was a right hemiparesis. Lumbar puncture findings showed normal pressure, 27 mononuclear cells and a protein of 75 Mg.%. Within 3 weeks, during which time she was treated with penicillin, her fever settled, her behaviour became more normal and her papilloedema disappeared. Her lumbar puncture findings returned to normal and her hemiparesis became less marked. It was considered that she had suffered from septic cerebral /

cerebral thrombophlebitis possibly with involvement of the left lateral and the longitudinal sinus. The other possible diagnosis in this case is of acute disseminated encephalomyelitis, but the rather gradual onset of the neurological disorder, the presence of papilloedema, the focal fit and the suggestive evidence of pyogenic infection which responded to penicillin makes infective intracranial thrombosis more likely.

In all these cases (of which Cases 50 and 144 may be regarded as typical), as in those of Byers and Hass, the initial clinical picture was that of the preceding illness, and only when convulsions or the hemiplegia itself appeared did a suspicion of underlying cerebral damage arise. But this is not the invariable course in cases of intracranial thrombosis in childhood, and the following case (Case 169), in which there was pneumococcal septicaemia, and Cases 4, 85, 175 and 111 illustrate a more insidious onset of hemiplegia in which symptoms referable to the nervous system were prominent from the first few days of the illness. The frequent occurrence of venous and arterial lesions in the course of pneumococcal meningitis has been commented on by Cairns and Russell (1946). In fact there is some evidence to suggest that the major damage to the brain in these cases results from septic venous thrombosis and consequent cerebral infarction, and that the meningitis is a secondary development consequent upon the venous lesions. In Case 169 the lack of any history suggesting the sudden onset of hemiplegia argues in favour of venous thrombosis rather than arterial embolism.

There /

There remain four cases to be discussed - Cases 4, 85, 175 and 111. In two of these patients there was pyrexia of unknown origin for a period before the onset of hemiplegia. In Case 85 the boy was 5 years old at the onset of a sudden weakness of the left side without loss but with some impairment of consciousness. There was no preceding fever or illness of any kind. He was admitted to hospital where a left hemiplegia was noted but no abnormal lumbar puncture or haematological findings detected. He rapidly regained considerable function in the affected limbs and was discharged within two weeks of admission. The acute onset of weakness in this case might be taken to indicate the probability of an arterial embolism or thrombosis, but the very rapid improvement in his hemiplegia suggests that the original episode may have been a mild epileptic seizure more likely to have been the result of a venous lesion.

In Case 175 there was intermittent low grade pyrexia especially at night for two or three weeks before the boy, then aged 10 months, was noted to drag the left leg when crawling, though the arm was normal. About two weeks later he began to drag the left arm too for a period of two or three weeks, but thereafter the weakness in both arm and leg tended to improve, though persisting to a greater extent in the lower limb than the upper. The slow onset of focal weakness which spread in the course of weeks to involve the whole side, whilst the child showed low grade fever, argues strongly in favour of a focal cerebral thrombophlebitis of a type rather more common in the adult than the child. But unfortunately

no /

no detailed neurological examinations or spinal fluid investigations were made at the time. A rather similar story of gradual loss of full use of the left limbs in present in Case 111, though in this patient there was no history suggesting previous infection or being "off colour".

In Case 4 the child had never been well from the day following his third injection of mixed pertussis and diphtheria vaccine. He was irritable, fretful and lost his appetite. He was fevered for 8 weeks and lost weight, and then within 3 days gradually developed a paresis in the right upper limb and then in the right lower, without convulsions or any other apparent exacerbation in the generalised systemic upset. He was admitted to hospital for 4 weeks by which time his quite severe hemiplegia had already shown marked improvement. No abnormality of the cerebrospinal fluid had been found. The lapse of time between the onset of the immunisation and the onset of the paresis, together with the subacute onset of the neurological symptoms make it most unlikely that acute disseminated encephalomyelitis was the cause of this child's hemiplegia, and the gradual spread of the paresis over a period of 3 days can best be explained on the basis of spreading venous thrombosis or thrombophlebitis.

Acute Disseminated Encephalomyelitis. As in the case of intracranial thrombosis, the diagnosis of acute disseminated encephalomyelitis had to be made retrospectively on the basis of circumstantial evidence, particularly on the basis of whether any preceding infection was of the type frequently complicated by this condition. (Table 53)

In /

TABLE 53.

Acute disseminated encephalomyelitis as a cause of acquired hemiplegia

Case No.	Age	Presenting illness.	Onset of neurological abnormalities.	Course and treatment.
184	14 months.	Measles and bronchopneumonia.	Convulsions and right hemiparesis within week of appearance of rash.	Gradual partial recovery.
191	5 years.	Measles.	Drowsiness and head retraction 6 days after appearance of rash.	Gradual recovery. Hemiplegia noted after few days when drowsiness lessened.
20	31 months.	None.	Sudden generalised convulsions lasting 12 hours. Then severe coma for 2 days.	Noted to have hemiplegia after fit.
48	18½ weeks.	Vaccinated between 15-18 days before onset.	Generalised convulsion and apnoea aged 18½ weeks.	Generalised convulsions for 24 hours, 8 hours after 1st. fit. Hemiplegia then appeared.
94	41 months.	Diarrhoea and vomiting aged 35 months with convulsions.	Severe persistent generalised convulsions 6 weeks after initial fit having been well in meantime.	Treated with antibiotics and heavy sedation. Gradual recovery after being semi conscious for 4 days.

In Cases 184 and 191, in which acute neurological symptoms appeared respectively towards the end of the first week after a measles rash had appeared, and six days after the rash appeared, there seems little doubt about the diagnosis. The course is typical of that of "measles encephalitis" (Case 191).

In Case 48 the infant was found face down in his pram at the age of 18 weeks apparently dead, but revived on artificial respiration. It seems almost certain that he had had a fit and fits recurred within the next 24 hours, after which time a right hemiplegia was noted. The period of between 15 and 18 days which had elapsed after vaccination is longer than is reported in most series of cases of diffuse encephalomyelitis complicating vaccination, but in the absence of any other evidence suggesting prior infection or birth injury vaccination appears to be a probable cause of the cerebral condition in this patient.

In neither Case 20, nor in Case 94, was there any suggestion of immediately preceding infection to account for the onset of severe generalised convulsions and loss of consciousness, though in the latter there had been a convulsion associated with diarrhoea and vomiting 2 months before. Case 20 is an example of the sudden onset of convulsions of this type. There were normal findings in the spinal fluid in Case 20, but in Case 94 the Lange Gold curve was 00123310000 with 9 mononuclear cells and normal pressure. After periods of coma lasting 2 days in Case 20 and 3 days in Case 94 both children made a good recovery but both showed hemiplegia and, most /

most interestingly, evidence of lower motor neurone lesions. In Case 20 the left leg showed definite weakness and some wasting below the knee, whilst in Case 94 there was a left third nerve paresis and marked peroneal wasting in the affected leg. The latter findings, indicating the probability of cord involvement suggest that diffuse encephalomyelitis without evidence of prior infection was the probable diagnosis in both cases, though it is impossible to be dogmatic about the diagnosis in either.

Probable "Toxic Encephalopathy". In order to avoid a discussion as to whether whooping cough encephalitis is an entity (Miller and Stanton, 1956) or the result of relatively non-specific changes found in many infective conditions (Dolgopol, 1941), it is necessary to interpret the term "Toxic encephalopathy" broadly. But it is convenient to do so, for the pathological changes described in children dying as a result of pertussis (commonly with convulsions) vary greatly and the clinical course is remarkably similar to that of patients dying as a result of "Toxic encephalopathy" complicating other conditions.

Two of the 3 cases which have already been described (Cases 19 and 196) have a fairly typical history of severe whooping cough being complicated by convulsions or the appearance of hemiplegia between the third and sixth week of the illness.

In Case 69 the history is not so typical, for the whooping cough was mild, complicated by severe chronic tonsillitis, and convulsions occurred within the first week of the illness.

It /

It is possible that this child may have suffered from cerebral thrombophlebitis, and the protracted period for which convulsions were present before hemiparesis was noted, is in favour of the diagnosis. However, closer inspection of the case notes reveals that the hemiparesis was noted immediately the child got out of bed for the first time, and it is probable that it had been present considerably before it was noted.

Toxic encephalopathy was considered to have complicated all the three patients in which hemiplegia occurred in the course of pneumonia. In all three cases the pneumonia was severe and accompanied by marked cyanosis and respiratory distress necessitating the use of oxygen. In all three the convulsions occurred on the first or second day of the pneumonia (though Case 6 had had a cold for a week before this). The convulsions were very severe in all three cases and were immediately followed by hemiparesis, which was rather slow to improve in all. This course of events is very similar to that described in similar cases by Marie (1888). Whether the real physio-pathological explanation of the lesions in such cases of severe pneumonia is that they are the result of severe persistent cerebral hypoxia or not is still debated (Chornyak, 1934).

Cerebral embolism and septic cerebral thrombophlebitis may both occur in the course of pneumonia, especially in infancy, but most of the cases in which these complications have been reported had been ill for some weeks before cerebral complications were present.

Trauma. /

Trauma. (Table 54) In 4 patients acquired hemiplegia resulted from trauma. In Cases 15, 165 and 195 this was directly to the skull but a fracture is known to have been present only in the latter, and only in this case was the patient unconscious. As in the majority of cases of head injury complicated by hemiplegia the underlying pathology is obscure, localised intracerebral haemorrhage, direct cerebral laceration or secondary hypoxic effects of the cerebral cells themselves all being possibilities.

In Case 15 the history of the limbs on one side being flaccid within minutes after a fall at the age of 10 months is unusual, but well authenticated. It seems possible that a brief epileptic fit may have occurred as a cause of, or immediately after the fall, for the child was found only after she had actually fallen. Again the underlying pathology remains unknown.

In Case 165, the child fell from a refugee cart whilst the family were escaping and was drowsy, lethargic, irritable and anorexic thereafter. Hemiplegia was noted only some weeks after the episode, but the child was never well during this time, and since only very scanty medical attention was available, no investigations are available to assist in determining if a subdural haemorrhage or fractured skull with secondary infection or direct cerebral laceration was present.

In Case 52 the child fell whilst running with a pencil in the mouth and perforated the left tonsillar bed and carotid artery with the point. She immediately had a generalised convulsion and thereafter showed a right hemiplegia. In the majority /

Trauma as a cause of acquired hemiplegia

Case No.	Age	Accident	Onset of neurological abnormality.	Course and treatment
15	10 months.	Fall from pram.	Face twisted and flaccid left limbs noted immediately after.	Some improvement within few days.
52	4 years.	Fell and penetrated left tonsillar bed and carotid artery with a pencil.	Immediate generalised convulsion with loss of consciousness and then right hemiplegia.	Some gradual improvement within about one week, but persistent right hemiplegia.
165	4 months.	Fell from cart on to head.	Irritability. Drowsiness and refusal to feed next day.	Gradual improvement in a few weeks, but persistent hemiparesis.
195	3 years.	Fell from air raid shelter. Fractured skull.	Loss of consciousness for some hours. Hemiplegia noted immediately after.	Observed for few days in hospital for fractured skull.

majority of similar cases thrombosis in the carotid artery can be demonstrated, but this may take some time to develop and the hemiplegia consequent upon it may not be evident for some hours (Braudo, 1956).

Probable Cerebral Embolism. One boy developed a left hemiparesis in the course of very severe diphtheria, complicated by endocarditis and palatal paresis at the age of 21 months (Case 129). Though "toxic encephalopathy" as a cause of the hemiplegia cannot be excluded, the severity of the hemiplegia with only slight recovery in the presence of endocarditis is much more in favour of an arterial lesion. Certainly the history is very similar to that in a number of the cases of Rolleston (1912).

Unknown Pathology. Case 121 is presented in toto for it is impossible to discuss her case without giving the full clinical details.

Case 121.

> Right hemiplegia of moderately severe degree, epilepsy and overactivity after convulsions. The only child born to a healthy mother and father. Mother aged 20 at time of birth, had been an acrobatic dancer. Both parents right handed.
>
> The child was born at home after a normal pregnancy. Delivery was spontaneous at term after a labour of 24 hours. The birth weight was 7 lbs., and the child cried at once, seemed healthy. She was a rather sleepy baby during her first year but active enough when awake. Held her head up at the age of 2 months, sat with support at the age of 6 months, and at 51 weeks she got up and walked, never having crawled. She used both hands to play with toys from the age of 6 months. From the age of about 1 year there was a change in her behaviour. She became active, curious and wanted to explore everything. She was beginning to enjoy nursery rhymes and was called a chatter-box by her father by the age of 20 months.
>
> From /

From the age of 15 months she had generalised convulsions with loss of consciousness lasting for up to 3 hours on an average every 3 months. On 3 occasions admission to hospital was necessary, but on no occasion was neurological abnormality or abnormality of lumbar puncture noted. The fits occurred in spite of regular administration of phenobarbitone. They tended to occur when her grossly enlarged tonsils, which were associated with enlarged cervical glands, were acutely inflamed.

At the age of 30 months she had a convulsion which was associated with loss of consciousness for 2 hours, during which she had apparently had the greatest difficulty in breathing and been intensely cyanosed. She was admitted to hospital and immediately given oxygen and paraldehyde. With this treatment the generalised convulsions ceased but some twitching and loss of consciousness persisted until next day. For 4 days she showed fever, disorientation and semicoma, very variable in degree, generalised increase of muscle tone and a tendency to lie in opisthotonos. Lumbar puncture findings were normal. She showed generalised wave and spike forms in the EEG.

She remained in hospital for 2 months, during which time the generalised rigidity became less and a right visual field defect, right hemiplegia emerged. Her behaviour was erratic, grossly overactive and very confused. She showed no purposeful activity, tended to lie in the foetal, generally flexed position. The showed no comprehension of what was said to her but appreciated the sound of the voice and other noises. She was incapable of any speech but screamed when touched or excited. There was nystagmus on movements of both eyes in all directions. Right facial paresis was present. The movements of the left limbs were inco-ordinate to some extent but full. Those of the right were slight and voluntary movement was very slight except in response to painful stimulation. Any voluntary movement that was attempted with the right arm was accompanied by marked athetosis of the fingers, wrist and arm. The athetosis was rather more rapid than the typical athetosis of a hemiplegia and less rapid than true chorea. In its distribution it was similar to athetosis in that it was most marked in the fingers, but it also extended to the wrist, elbow and slightly in the right shoulder. There was a spastic increase of tone in the right arm and leg compared to the left, and the biceps, triceps, supinator, knee and ankle jerks were increased. The right Hoffmann was positive and the right plantar was extensor. The grasp and sucking reflexes were positive bilaterally. There was neglect of the right side but it was impossible /

impossible to date whether some loss of sensation was also present. Swallowing of solids was very difficult for her. Her eyesight appeared to be grossly defective. Her sleep was disturbed many times each night by constant screaming.

Two months after discharge from hospital she began to be interested in food and to have less difficulty swallowing. At the same time she began to use the left hand for playing with objects. Three months after discharge she was able to sit with support and shortly afterwards without support.

Six months after discharge from hospital, as her vision appeared to improve she was able to stand, first with support and then without. At about the same time she began to use the right hand a little and learnt to say "Mum" and "Dad". She was very unsteady when she first got on to her feet again, but had become steadily less so. She had become progressively more overactive, was never still, touching and moving everything and putting much of what she handled to her mouth. She showed a persistent tendency to go into things on her right hand side.

When she first came home the parents noted that the least sound was inclined to make the child double up, head on chest, lose or partially lose consciousness for about a few seconds and then appear as usual once again. These attacks occurred as often as 15 - 20 times a day initially but gradually became less frequent as time went on, and 6 months after her discharge from hospital they occurred only on much greater stimulation and only 2 or 3 times a day. The attacks also seemed less severe and amount to little more than momentary head nodding.

Examination one year after last hospital admission.
Height - 38½". H.C. - 19¼".

There was ⅛" shortening in the right arm, and ¼" in the right leg compared to the left.

The child was very disordered, wandering about touching and retouching objects in a meaningless fashion, never still and apparently completely unable to concentrate. Her interest could not be fixed for more than a moment on any given object. She seemed to comprehend simple commands from her mother. She could say "Mum" and "Dad" but had no other words. She used these words out of context, and one had the feeling that a degree of dysphasia was present.

There was a right visual field defect but the degree of this could not be accurately assessed.

A right facial paresis of upper motor neurone type was evident. Swallowing was infrequent and there was a tendency to drool.

The left arm showed a degree of athetosis of the /

the fingers on voluntary movement. The right arm and leg tended to be neglected and the arm very little used except to assist the left by staedying. The voluntary movements in the right arm were accompanied by gross athetosis of the fingers and wrist, which had changed in character to a typical slow writhing type. Fine and rapid finger movements were impossible. The right arm tended to be held in the position of flexion at the elbow, adduction at the shoulder and flexion at the wrist and fingers. The forearm was pronated. The position was accentuated when she walked or used the left arm.

Her gait was unsteady and she leant forward acutely as she proceeded, the right leg swinging outwards, the foot in a position of equinus and the toes dragged. The tone of the right limbs was much increased and was spastic in type, more increased in the right than the left. The biceps, triceps, supinator, knee and ankle jerks were increased on the right compared to the left. The jerks in the left arm were somewhat brisker than normal and there was some increase of tone of spastic type. This was more evident in the left arm than leg.

The right Hoffmann sign was positive, the left negative. The right plantar was extensor, the left flexor.

Sensory findings were probably normal but not fully testable.

The right hand and foot were colder than the left.

A number of possible diagnoses to account for the symptoms, neurological signs and course of this patient are possible. The history of a change in behaviour at the age of one year followed 3 months later by the beginning of epileptic attacks strongly suggests a slow degenerative process in the brain, and the presence of myoclonic jerks later, in association with a hemiplegia and residual epilepsy, is also compatible with this. On the other hand there was no increase in the spinal fluid protein at any time, and when last seen the child was improving rather than deteriorating.

Another /

Another possibility is that the change in behaviour and the fit three months later represent a low grade inflammatory or thrombotic process in the cerebrum, possibly thrombophlebitis, and the evident severe neurological deterioration after the last fit was the result of hypoxic damage occurring during it.

Still another possibility is that the last convulsion and the severe neurological deterioration which followed it represent an acute disseminated encephalomyelitis complicating an upper respiratory tract infection in a child previously predisposed to epilepsy.

Certainly this history serves to illustrate the difficulties of attempting to make a clinical diagnosis of the various encephalitis and encephalopathic syndromes in childhood. It is reassuring to find that these difficulties have been repeatedly mentioned by recent authors, (Van Bogaert, 1954; Brewis, 1954; Radermecker, 1956).

Chapter 4d.

Aetiological Studies of Hemiplegia of Unknown Origin.

It proved impossible to classify twelve patients as suffering from either congenital or acquired hemiplegia. In three of them the mothers were definitely mentally ret- retarded and one was very emotionally disturbed. In these the failure to classify may have been the result of the rather inadequate histories which were obtained. But in the remaining nine patients the case histories were felt to be reasonably full, and details of the labour and deliveries were obtained from the maternity

maternity hospitals in 8 of the cases and later hospital notes being available in 10.

There was no clear cut history suggesting significant neurological abnormality immediately after delivery, or within a few days of birth in these patients, except for some feeding difficulty in cases 88 and 113, nor was there any history to suggest a more or less marked change in activity with the onset of hemiplegia after this period. The case histories are summarised in Table 55.

It will be seen that 4 patients were delivered after abnormal parturition (Cases 36, 160, 168, 88). In Case 160 the mother suffered from tuberculosis and was never well during her twin pregnancy. In Case 36 the mother fainted and was unconscious for some minutes at the seventh month of gestation. In Case 168 occasional labour pains were felt in the last month prior to the onset of labour proper. In Case 88 the mother fell heavily at 7½ months' gestation. In no case was labour or delivery abnormal, and all the infants breathed immediately after delivery, in contrast to the patients with congenital hemiplegia. The majority of the babies appeared to be normal immediately after birth, but strabismus was noted in Cases 58 and 113 shortly after, and hypomandibulosis was present in Case 113.

In 6 cases retarded motor development first gave rise to parental concern (Cases 36, 58, 119, 130, 168 and 194). In 3 cases other neurological abnormalities attracted the first concern. In Case 134 the child was observed to move the left limbs less than the right at the age of about 4 months, and /

and at the age of about 5 months the district nurse noted hydrocephalus which subsequently became arrested. Case 190 was noted to move the left hand less than the right when between 1 and 3 months old. In Case 88 the head was held tilted to the left at the age of a few weeks. In 3 cases (Cases 160, 199 and 204) it was impossible to obtain a detailed history, and the time at which the hemiplegia was first observed is unknown.

In the other cases the hemiplegia was noted to be present before the age of 3 months in two (Cases 58 and 88), in three by the age of 4 months (Cases 119, 134, 190), in two at the age of about 6 months (Cases 36 and 113) and in Case 130 at about the age of 15 months. In Case 168 it was apparent between the ages of 15 and 30 months.

The history of a hemiplegia being observed before 6 months in the majority of patients, usually when the child began to use the arms, is quite compatible with that of congenital hemiplegia, but the absence of a history suggestive of birth trauma, or hypoxia and the apparent normality of most patients in the neonatal period is against this being due to abnormal birth.(Norman, 1947). Though it is possible in Case 168 that the hemiplegia was present immediately after the convulsions at the age of 15 months, in the remaining cases there is no history suggestive of the acute onset of hemiplegia as the result of acquired disease. There seem to be three possible explanations for these cases. Firstly the histories may be more inadequate than is thought, and pregnancy and labour may have been more disordered than is apparent, or acquired diseases /

diseases may not have been observed.

Secondly there may have been an insidious onset of hemiplegia, commonly at the age of a few months, in these patients without symptoms of generalised disturbance. As will be described, in a proportion of cases of acquired hemiplegia, usually rather older children, the onset of hemiplegia may be gradual and accompanied by little systemic upset.

Thirdly these cases may be cases of congenital hemiplegia in which parturition was relatively uncomplicated and in which the condition was not due to birth injury but to other factors, possibly developmental abnormalities in earlier pregnancy. A few pointers suggest that this may be the most likely explanation in the majority of the cases. The appearance of hemiplegia before the age of 6 months in at least 7 cases, the occurrence of retarded development in at least 7 cases noted before the age of 6 months, and the fact that 8 of the patients were found on examination to be mentally defective suggest an onset before or at the time of birth or very shortly after. The occurrence of hypomandibulosis in Case 113 and of hydrocephalus believed to be of congenital origin in Case 134 means that at least 2 patients had associated congenital anomalies. The ages of the mothers (4 of the 11 whose ages were known were over the age of 39), their relatively low fertility and the high rate of miscarriages in other pregnancies would be compatible with the suggestion that these cases of hemiplegia are due to developmental anomalies rather than to birth injury.

It is interesting to compare this series of patients with that /

that presented by Yannet (1949) in which exactly the same problem arose. A proportion of patients with hemiplegia apparent in the first year of life and probably congenital had no history suggestive of birth injury. Though he admitted that this might be due to defective records in some of the patients, he felt that the majority were probably due to the effects of genetic factors and other causes of developmental abnormalities in early pregnancy. These factors were probably responsible for about one quarter of his cases, a higher proportion of whom were mentally defective than in this series since they were largely derived from institutions.

On the other hand it seems undesirable to classify patients in this category as congenital dogmatically, when there is no evidence of their showing definite neurological abnormalities in the neonatal period. "Probably congenital" is probably a better description, and "of unknown aetiology" seems the most correct designation of all.

TABLE 55.
Aetiological factors in hemiplegia of unknown origin.

Case No.	Sex	Place in family.	Age of mother.	Pregnancy.	Delivery	Neonatal period.	First sign of neurological abnormality.	Age when hemiplegia noted.	Findings on examination		Stillbirths & neonatal deaths.
36	M.	2/4	24	Fainted and convulsions for some mins. at 7th. month.	Spont. vertex after 4 hours.	Normal.	Noted to be moving less than other children ated 4 mths. Retarded milestones.	Shortly after age of six months.	Mod.sev.Rt. hemiplegia. Epilepsy. Mental Impairment. Strabismus.	Sib.died of hydrocephalus aged 4 months.	-
58	F.	4/4	28	Normal.	Spont. vertex after 9 hours.	Normal.	Squint noted after birth. Retarded milestones.	2-3 mths. when noted not to be moving Rt.hand.	Mod.sev.Rt. hemiplegia. Mental deficiency. Epilepsy.	Both parents of low intelligence.	-
119	M.	2/2	28	Vague ill-health. Neurotic.	Spont. vertex after 12 hours.	Normal.	All milestones retarded. Walked aged 2.	Used only right hand from 4 mths.	Mild left hemiplegia and mental defect.	Mother paranoid. Father alcoholic.	-
130	F.	1/1	21	Normal.	Spont. vertex after 8 hours.	Normal.	All milestones delayed.	Reached with Rt. hand aged 15 mths. but never left.	Severe left hemiplegia. Mental defect. Strabismus. Congenital dislocation of of left hip.	Negative.	-
134	M.	12/13	41	Normal.	Spont. vertex after 8 hours.	Normal.	Hemiplegia noted at 4 mths. with head enlargement.	4 months.	Severe left hemiplegia. Arrested hydrocephalus.	Sib. died in convulsions aged 3. No known cause.	10th.& 11th. children twins, and 2nd. died after prem. breech extract.
160	F.	2/3	41	Twin pregnancy. Pulmonary tuberculosis.	Unknown.	Unknown.	Unknown.	Before the age of 3 years.	Mod. severe right hemiplegia. Mental defect.	Mother of low intelligence.	-
168	M.	1/1	31	Normal apart from occasional pains in last week of pregnancy.	Spont. vertex after 4 hours.	Normal.	Milestones retarded. Convulsion aged 15 mths. Meningitis aged 3.	Unknown.	Mild right hemiplegia. Severe mental defect.Epilepsy.	-	-
190	M.	1/2	22	Normal.	Spont. vertex after 13 hours.	Normal.	Noted not to move Lt.hand aged 3 mths. and fingers found to be stiff.	4 months.	Mod. severe right hemiplegia with athetosis.	-	-
199	M.	2/4	37	Normal.	Spont. vertex after 4 hours.	Normal.	Retarded speech development and unsteady on feet aged 20 months.	Unknown.	Slight right hemiplegia. Mental defect.	One sib. diplegia. One sib. backward.	-
204	M.	3/3	Unknown	Unknown.	Unknown.	Unknown.	Unknown.	Unknown.	Mild left hemiplegia. Mental defect.	Father mentally retarded and alcoholic. Mother mentally retarded.	-
88	M.	1/2	39	Heavy fall at 7½ mths.	Spont. vertex after 12 hours.	Diff. in swallowing.Slow feeding.	By age of 2 weeks seemed to have head tilted to left. Slow milestones.	By age of 6 weeks.	Mod. severe left hemiplegia.	-	-
113	M.	1/1	34	Normal.	Spont. vertex after 20 hrs.	Normal except for slow feeding.	Squint of left eye noted at few weeks. Slow to sit up.	Slow to use right hand aged 6 - 7 months.	Severe right hemiplegia. Hypomandibulosis.	-	-

CLINICAL FINDINGS IN HEMIPLEGIA.

In Table 56 are shown some of the more important findings in cases of congenital and acquired hemiplegia. It will be seen that apart from the fact that a higher proportion of patients with acquired hemiplegia were aphasic and rather more epileptic, there are no marked differences in the prevalence of clinical findings in the two groups. There is some justification, therefore, for considering the clinical findings of all 75 hemiplegic patients together, rather than considering the congenital and acquired cases separately.

Side of the Paresis.

The side of the paresis is of importance from two points of view. Firstly this is essentially a right handed culture, and the majority of people have a natural right handed preference. A left handed person is somewhat handicapped, and a child who has to use the left hand when naturally right handed is at a further disadvantage, especially if he has had to change in later childhood (Hildreth, 1948).

Secondly, when the master or dominant side is affected, most commonly the right, aphasia, certain reading and writing difficulties and possibly behaviour disturbances occur more frequently. Table 57 shows that whilst the severity of the paresis assessed clinically was not more severe if the right side was affected, certain other associated abnormalities were more frequent. It is interesting that only one patient with aphasia complicating a left hemiplegia was found, and he was left handed before the onset of his disease and was still

TABLE 55

Comparison in the findings in congenital and acquired hemiplegia in childhood

Origin of hemiplegia	Male	Female	Rt.	Left	Totals	Severity of hemiplegia			More than 1" shortening in affected upper limb.	Marked Athetosis	Severe trophic changes	Epilepsy	Asphasia
						Mild	Moderately severe	Severe					
Congenital	20	10	20	10	30	11	8	11	13	9	6	7	2
Unknown	9	3	7	5	12	5	5	2	2	2	1	2	1
Acquired	23	10	17	16	33	11	13	9	11	8	5	15	9
Totals	52	23	44	31		27	26	22	26	19	12	24	12

left visual fielded afterwards.

In most surveys hemiplegia is said to occur with equal frequency on the right and left sides or to be rather commoner on the right (Osler, 1889; Freud and Rie, 1891). In a recent series of 173 cases of hemiplegia, 56% were on the right side, compared to approximately 59% in the present series (Hood and Perlstein, 1955).

The Severity of the Hemiplegia.

The criteria on which the severity of hemiplegia was judged has been described in the section on classification. It must be stressed, however, that the degree of functional loss is not necessarily proportional to the severity of the paresis. Loss of function depends upon a large number of different factors, including the severity of the paresis, the degree of dwarfing, the presence or absence of involuntary movements and sensory loss, the severity of trophic changes and to a degree the intelligence of the child and his ability to compensate for his neurological deficits.

In general, mild cases were able to use the affected upper limbs independently but clumsily. There was impairment of power and coordination of fine and rapid movements of the fingers, and supination of the forearm was weak and sometimes slightly restricted. Contractures tended to be slight and trophic changes mild. Associated movements of flexion were almost invariably present in both the affected limbs, the tonic reflex was accentuated on the affected side, the grasp reflex was normally present and jerks were markedly increased.

Sensory /

TABLE 57

The findings in 75 cases of hemiplegia by sex and the side of the body affected.

			Severity													
			Mild	Mod.	Severe	Total	Epil-epsy.	Normal	P.H.	M.H.	Ineduc-able	Not yet at school	Aphasia	Speech defect	Visual defect	Overactive behaviour
Male	Rt.	31	12	10	9	52	8	7	5	5	5	9	11	9	6	4
	L.	21	8	8	5		6	9	5	4	1	2	1	5	1	3
Female	Rt.	13	4	4	5	23	5	3	2	2	2	4	0	4	1	2
	L.	10	3	4	3		5	4	2	1	1	2	0	3	3	0
		75	27	26	22	75	24	23	14	12	9	17	12	21	11	9

P.H. = Physically handicapped.
M.H. = Mentally handicapped.
Mod. 2 Moderate.

Sensory changes were usually absent or very slight. The Hoffman sign might or might not be present, but the plantar response was invariably extensor.

Moderately severe cases used the affected arm only as an assistant, and neglect of the limbs was probably the most striking clinical feature in many patients. Handling was clumsier than in the mild group, limitation of extension of the fingers, abduction of the thumb, supination of the forearm, and sometimes of extension at the wrist all being present. Fine and rapid movements of the fingers were very much impaired. There was often difficulty in releasing the grasp. Contracture was more frequent and more severe, and sensory deficits, trophic changes, dwarfing and involuntary movements of athetoid type were more in evidence. Whereas very few of the patients in the mild group had had to have tendo Achilles lengthenings, about half those in the moderately severe group had had them.

Severe cases were those in which the upper limb was so affected that, for practical purposes, the function was confined to giving some support to the useful arm. The hand was usually used very little, and objects had to be held with the side of the hand against the trunk on account of the inability to extend the fingers and the tendency for there to be fixed flexion contractures of fingers, wrist and elbow, and pronation contracture of the forearm. Involuntary movements were marked unless restricted to an extent by contracture. In the lower limbs flexion contractures at the hip often occurred, and together with the contractures at the knee and shortening of the tendo Achilles served to make walking very difficult. /

difficult. Surgery had often been repeated in efforts to alleviate the contractures of the tendo Achilles and pronators of the forearm, but with little success in the majority unless accompanied by persistent splinting.

Though there was a tendency for dwarfing, vasomotor and sensory changes and the involuntary movements to parallel the paresis in severity, this was not a consistent finding, and a few cases with mild paresis were placed in the group of moderately severe hemiplegia on account of the effects of involuntary movement or of sensory deficits on upper limb function.

The relative severity of loss of function in the upper and lower limbs.

In general the paresis of the upper and lower limbs was proportionate, the arm being more affected than the leg. But this was not necessarily true of limb function, for the effects of involuntary movement and sensory loss were much more apparent in the upper limb whilst dwarfing was more of a handicap in the lower limb. In a few cases the arm was much more severely paretic, dwarfed and affected by trophic changes than would be expected, and in a single case the leg was more severely involved than the arm. In Case 11 the lower limb was particularly severely involved.

Impairment of voluntary movements.

The impairment of voluntary movements which occurs in hemiplegia has been much studied since the work of Beever (1903), Sherrington (1904) and Walshe (1923, 1929). It has become increasingly apparent that it is patterns of movement which /

Case 11.

Hemiplegia affecting the lower limb relatively severely.

Boy aged 13 at the time of examination who slowly developed weakness of the left upper and lower limbs between the ages of 12 and 15 months.

He was the first of three children born to healthy parents after uncomplicated parturition. His birth weight was 8½ pounds. His development during infancy was uncomplicated and he was standing without support and saying his first words at the age of 12 months.

At this time the mother observed that his left hand seemed clumsy in play and after some days noted that it felt "kind of stiff and lifeless" when she washed it. Within ten or fourteen days he began to drag the left leg when crawling. Both limbs became progressively though not markedly weak. When he walked at the age of 15 months without support he dragged the left leg a little and limped towards that side. After this time the strength and function of both arm and leg seemed to improve, so that clumsiness of the hand was noted only when he was making precise small range movements. He was able to play successfully with other children but could not run as quickly as they could. He was referred to hospital at the age of 4 years on account of a claw foot which improved to some extent with physiotherapy. He was doing well at a normal school at the time of examination.

On examination he was a well built boy of average height and intelligence whose left upper limb was 1½" shorter and left lower limb 1" shorter than those on the right. His speech was normal. He was right handed.

There was a slight left facial paresis. There was slight impairment of power on voluntary movements in the left limbs compared to the right which were normal. Weakness was most marked distally and affected extension, abduction and adduction of the digits, supination of the forearm, movements of the toes and dorsiflexion of the foot more severely than movements of proximal parts. Movements in the upper limb were full in range but he could not dorsiflex the left foot to within ten inches of a right angle. He was able to handle objects remarkably well with the left hand, however, and used it for feeding and as an assistant to the right whilst dressing though he could not manage buttons left handed. Slight athetoid movements of the left hand were noted when he used it. He could not hyperextend the left hip nor fully abduct it. Knee movements were full in range.

There /

There was an increase of tone of spastic type in the left limbs, the tone of the right being normal. The biceps, triceps, supinator jerks were moderately increased. The knee and ankle jerks were markedly increased and left ankle clonus was present. The left plantar response was extensor, the right flexor.

When he walked he did so on the left toe with the foot clawed and swung the leg in a wide arc from the hip, with the knee slightly flexed. He limped towards the left side, and when he tried to run his gait became very ungainly and he was liable to fall on rough ground.

No abnormalities of sensation were observed. The left hand and foot were colder than those on the right, the foot being colder than the hand. There was wasting of the muscles of both the left arm and leg compared to the right, the leg being relatively more severely affected.

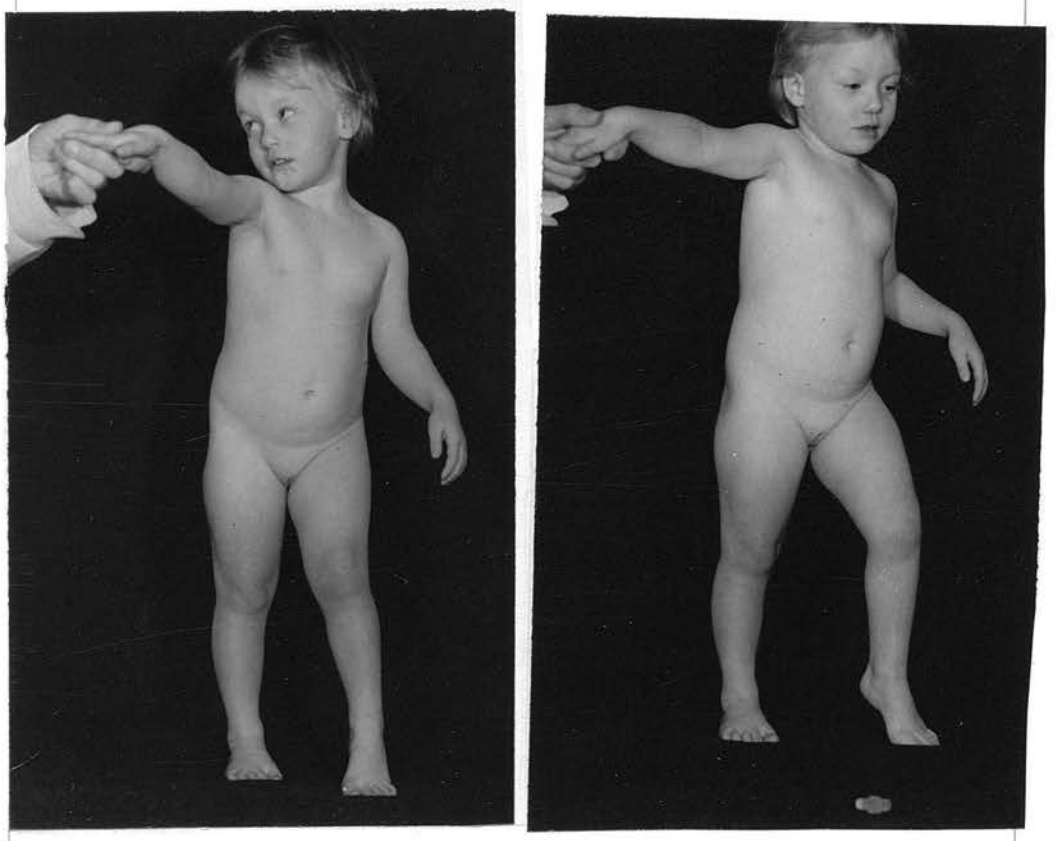
which are lost and often replaced by more primitive movement patterns, rather than that individual muscles are paralysed or weakened. In particular fine and rapid movements of the fingers which are acquired relatively late in infancy and early childhood are prominently impaired. It is very difficult by clinical testing to judge how much of the impairment of voluntary function is due to actual weakness of movement and how much to incoordination of movement. For example, some children by skilful utilisation of the grasp reflex appeared to hold objects wery strongly, though examination would reveal that voluntary finger flexion was poorly coordinated and weak. One boy working as a cooper used to hold wood for sawing in his affected hand by this means, though voluntary extension and flexion were severely limited. In all cases, however, it was fine finger and thumb movements which were most impaired, extension and abduction being the first to be affected. Alternating pronation and supination of the forearm could not be performed rapidly in most cases and the power of supination was almost invariably diminished. Wrist extension, elbow extension and shoulder abduction were the other movements impaired in order of the frequency and severity of their being affected. In the lower limb movements of the toes were always impaired, then rotatory movements at the tarsus and ankle, then abduction and extension at the hip, extension at the knee, and only rarely was hip flexion affected.

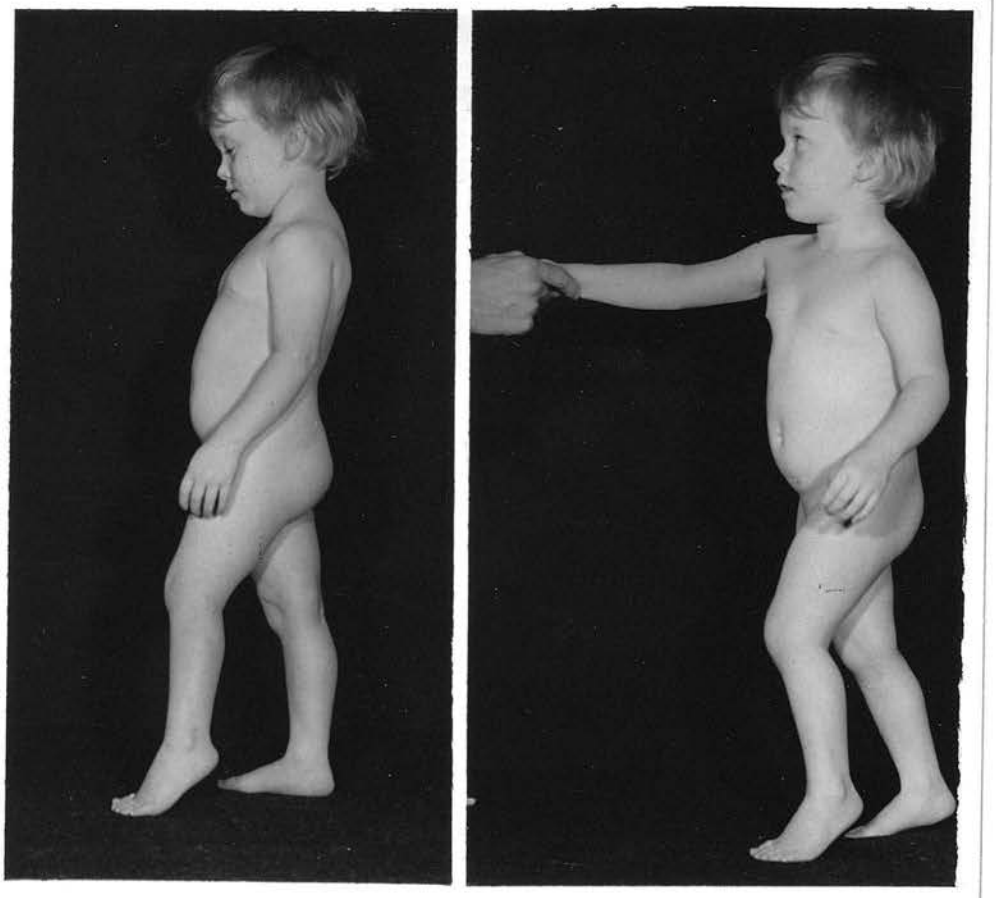
Spasticity.

The severity of spasticity and the degree of exaggeration of /

Figure 2.

Congenital left hemiplegia in a prematurely born twin. The characteristic attitude of the affected limbs during walking is well shown.





of stretch responses were usually roughly proportional to the severity of the paresis, but the degree of exaggeration of the tendon jerks was not. This was because the contracture in severe cases of hemiplegia was often so severe that tendon jerks which would otherwise have been very brisk were limited. The occasional finding of an increase in the tendon jerks in the contralateral leg, especially in mentally defective patients is worth noting.

Other reflexes.

The tonic neck reflex was almost invariably present even in mild cases of hemiplegia though in cases showing severe contracture of the affected limbs the full movements might not be observed.

The plantar responses were invariably extensor on the affected side, though in a proportion of patients it took some time to convince oneself that this was so owing to withdrawal of the limb and equivocal movements of the toes. No cases were found in which the plantar response in the contralateral leg was extensor, even when the tendon jerks were somewhat exaggerated, as has been described.

The Hoffman response was much less frequently present than the Babinski and was not found to be reliable, possibly because in many of the younger children the fingers were hyperextensible rather than contracted and it was difficult to provide the correct stimulation to elicit the response.

The grasp reflex was present in the majority of cases but unfortunately it was not closely studied. The author was largely ignorant of the component groping, grasp and traction /

traction response at the time of the survey (Denny Brown, 1957; Twitchell, 1958).

The sucking reflex was obtained relatively seldom in the waking state, but observation since the time of the survey has shown that in hemiplegic children under the age of 7 or 8 years it is nearly always present on the affected side when they are drowsy or half asleep.

Generalised associated movements.

In this series involuntary movements of the generalised associated type were apparent in every case of hemiplegia. The severity of the movements and the ease with which they were elicited depended to some extent on the severity of the paresis in the limbs. If the child exerted himself sufficiently, especially with the head turned to the hemiplegic side, so that the tonic neck reflex came into action, they could always be produced, even in the mildest of cases of hemiplegia. The existence of these movements was sometimes of diagnostic help.

The activities which tended to produce the generalised associated movements were those in which the child exerted himself to any considerable extent. In severe cases of hemiplegia they might be produced by acts as slight swallowing or by the child becoming excited. In slight cases of hemiplegia more strenuous activity might be required, running, lifting a weight with the unaffected arm with the head towards the hemiplegic side, or by means of performing the tonic neck reflex briskly. In most cases the associated movements were more easily produced with the child standing than /

than lying, with the head extended than flexed, and after he had had some exercise of the affected limbs.

The movements of the affected upper limbs were predominantly flexor in type in all but two of the cases of hemiplegia examined fully. When the child exerted himself he showed adduction at the shoulder, flexion at the elbow, wrist and finger, pronation of the forearm and adduction of the thumb. The findings in the lower limbs varied more. In most cases flexor positions were assumed, contrary to the usual statements of the findings in adults, the hip being adducted and the hip and knee flexed. The foot took up a position of plantar equinus so that the shortening of the limb resulting from hip and knee flexion was to some extent compensated. In 27 of the 74 cases examined in detail, extensor positions were assumed in the leg on exertion, sometimes with hyperextension at the knee. In most cases, therefore, exertion tended to precipitate an exaggeration of the position in which the limb was held as a result of contracture.

The tonic neck reflex in hemiplegic patients affords another clue as to the nature of the reflexes which result in the associated movements. In the vast majority of patients it was positive, but the movement it produced in the limbs varied very greatly. In those with marked contracture when the head was turned to the normal side, the tendency to extension of the affected arm was slight, but in most cases a very marked extension took place. In mild cases and those in whom contracture had been prevented the arm could frequently be made to take up a position of adduction and internal rotation /

rotation at the shoulder, extension at the elbow, pronation of the forearm and flexion of the wrist and fingers. This decerebrate like position of the arm was associated with a marked increase of tone in both affected limbs and always with a degree of extension in the affected leg, frequently with slight hip flexion.

The degree to which the limbs responded to the tonic neck reflex was dependent upon the severity of the contracture present in the limbs, but also upon whether the positions of associated movement in the limbs tended to be strongly flexor or not. Thus in the cases where running with the head held straight produced markedly flexor patterns in the paretic arm the tonic neck reflex usually produced only slight extension when the head was turned to the sound side. In the two cases in which normal exertion produced only extensor positions in the affected arms very little flexion was produced by turning the head towards the affected side.

The importance of the generalised associated movements from the point of view of the effects of their function is two-fold. In the first place in slight cases of hemiplegia acts requiring the use of two hands or of the paralysed hand when the child is walking or otherwise exerting himself are made difficult by the tendency for the affected limb to assume the position of predilection, usually the position of flexion. It is very noticeable that children taught shoemaking are more successful in schools where they do not have to work the sewing machine with their legs than where they do.

The generalised associated movements are important in the second /

second place because to some extent the child may be able to utilise them to improve his walking pattern and appearance.

Imitative movements.

Descriptions of "imitative movements" in the literature are very frequently in fact of movements of the generalised associated type, postural mechanisms whose true nature has not been recognised. True imitative movements are not nearly so common and were observed to some degree in only 15 cases in this series, 9 of them slight cases of hemiplegia and 4 moderately severe. The movements most commonly imitated are unfortunately those tending to place the limb in the postion of predilection, usually flexor positions. Imitative movements are more likely to occur in movements involving considerable exertion on the sound side. The simplest test is to ask the child rapidly to pronate and supinate the sound arm and watch the affected limb. It will usually be found that the generalised associated movement predominates and that a position of steady increased flexion and pronation is achieved but occasionally the wrist of the affected limb may be seen to extend and flex slightly. The affected limb also appears to be more likely to imitate movements arising as a result of emotion. Thus when the sound arm is suddenly raised in alarm to ward off a blow, the affected arm may follow suit.

Athetosis.

By athetosis is understood a condition in which involuntary movements of the affected limb, especially its distal parts /

parts, are accompanied by slow, inco-ordinate writhing movements of the fingers, and less commonly the wrist and the forearm. Similar phenomena may be observed in the toes and the feet in most cases.

Athetosis was of a rather similar pattern in most cases though the extent and severity to which the limbs were involved varied greatly in different cases. It was most commonly produced by movements of the affected arm which required a mixture of postural activity and voluntary muscular activity. The clumsiness of the affected hand in most cases of hemiplegia means that its activities are confined to grasping objects. Delicate manipulation is impossible.

Table 58

Severe Athetosis in Hemiplegia in Childhood

Severity of hemiplegia	No. of cases	No. showing severe athetosis
Mild	27	4
Moderately severe	26	5
Severe	22	10

It is most common when the child reaches for objects, and is maintaining the limb in a steady position prior to grasping the object with the hands, that athetosis is seen. Instead of a smooth rapid change in the position of the fingers from the more or less extended to the more or less flexed state, their movement is suddenly arrested, the tone of the whole limb increases momentarily, and the fingers then separate /

separate and extend, usually hyperextend slowly and irregularly. The movement is commonly most severe in the ring or middle fingers but varies in distribution. At the same time as the fingers extend and separate the wrist may extend, and even hyperextend in severe cases, and there is a tendency for the forearm to be pronated. After perhaps a quarter of a second or a second the fingers and wrist suddenly flex, sometimes with such violence that the hand is thrown from its carefully set position and the object is not grasped. The actual trigger mechanism of the athetosis seems to be the child's attempt to flex the fingers when the arm is being maintained in one position.

The extension and hyperextension found in the fingers during athetoid movements is usually in very marked contrast to the very limited range of finger extension which is possible by voluntary movement. Table 58 shows the number of cases whose athetosis was severe enough, in itself, to be a cause of limitation of function in the affected hand. In general, severe cases and moderately severe cases of hemiplegia were those which showed athetosis of the limbs of this degree. But a smaller proportion of slight cases did also.

Though it was a handicap in the majority of cases who showed it, athetosis could also be utilised by the child to increase the use of the affected hand. The cases in which athetosis occurred most frequently were those in which paresis was severe enough to have limited the power, accuracy and extent of voluntary finger extension more or less severely. Children so affected found that grasping objects /

Figure 3 .

Girl aged 4½ years with moderately severe acquired right hemiplegia, showing dwarfing of the right limbs with cyanosis of the right hand (a), deformity of the right hand, (b), and athetosis when she reached with the right hand, (c).

a.

c.

objects was difficult largely because they could not easily extend the fingers enough to grasp objects. Releasing objects was usually equally difficult. (Case 129).

By means of utilising the full extension of the fingers found in athetosis a number of children, quite severely handicapped by their hemiplegia, were able to open the hand enough to grasp objects and release them with surprising facility.

Choreoid movements.

Choreoid movements were much less frequently encountered than athetoid in hemiplegia in childhood, but a large number of cases in the earlier literature show that their occurrence cannot be considered rare. Cases of choreoid movements affecting the hemiplegic limbs were reported by Gowers (1874), Raymond (1876), Charcot (1887), Audry (1892) and Bouchaud (1894). Various transitional forms of disorder occur in which involuntary movements of athetoid as well as choreoid type may be found.

In type the choreoid movements that occur are interesting in that essentially they consist of a rapid intermittent series of involuntary movements which tend to bring the limb into one of the positions of prediliction. Usually the position eventually assumed is one of extension of the upper arm rather than flexion, but the position of the limbs fluctuates widely and markedly between the positions of flexion and extension when voluntary movement is attempted.

Even slight choreoid movements tend to handicap the function of the affected limb considerably because not only are /

Case 129.

Severe left hemiplegia with athetosis following diphtheria.

The first illegitimate child of mother aged 23 at the time of birth and unknown father. The mother later married and has three healthy children.

During her pregnancy she was well and the delivery was at term. Labour lasted 9 hours and the child was born spontaneously in hospital. Birth weight was 7 lbs. 5 ozs. The child cried at once and seemed normal. There was some vomiting at the 6th and 7th day with an elevation of temperature. The child became dehydrated and four subcutaneous infusions were given. Thereafter the neonatal course was normal. He sat with support at the age of 6 or 7 months, and walked and talked at the age of about one year. He used both hands when playing and there was no unusual unsteadiness or abnormality of the gait.

At the age of 21 months he developed diphtheria and was in hospital for four months, during one month of which he was extremely ill with endocarditis and palatal paresis. Because he was suspected of having meningitis at this time he had a lumbar puncture which gave normal findings.

On discharge from hospital at the age of 2 years it was noted that the child was unsteady on his feet, that he was walking on the left toes and that he dragged the left foot as he proceeded. The left arm was held bent across the chest and he used only the right hand. His speech had been normal and his pronunciation good before his hospital admission but was slurred and difficult to understand when he was discharged.

As time passed, the tendency to walk on the left toes appeared to become more marked and he had three tendon lengthening and Steindler's operations up to the age of 12 years, and as a result he now walks with the left heel on the ground. At the age of five he went to a normal school and managed to keep his place there until he left, though he was always in the bottom half of the class.

Examination. Height 64", head circumference 21". There was shortening of one quarter of an inch in the left leg and one third of an inch in the left arm compared to the right. Adolescent. Rather a dull boy of placid temperament. Replies to questions and responses to commands slow and unimaginative but in general accurate. Speech was normal. There was left facial paresis of moderate severity, upper motor neurone in type. Weakness of the left half of the tongue.

The /

The power and co-ordination of the left arm and leg was impaired. This was most marked in the left arm and especially in the hand and fingers. There was slight contracture of the right fingers in the flexed position, the right wrist and the forearm in the position of pronation. More marked than the contractures was the paralysis of voluntary movement. The fingers could not be extended more than a few degrees; the wrist could not be extended fully and there was great weakness of extension of the elbow. Supination was severely limited. Thus the movements of the left arm were very limited, but even these were complicated by very gross athetosis of the fingers, hand and wrist. On the least attempt to stretch the arm or pick up an object the fingers showed slowly writhing athetoid movements, becoming extended and separated, one from the other. The wrist hyperextended and the elbow extended more than could be achieved voluntarily. By utilising the extension athetosis of the fingers he was able to grasp objects. He could not release them once grasped, however.

The lower limb on the left showed great weakness and considerable muscular wasting, especially below the knee. The toes could not be moved. There was limitation of voluntary dorsiflexion at the ankle to just below the right angle. When the foot was moved the toes showed athetoid movements similar to those in the fingers.

The left limbs showed great spastic increase in tone compared to the right, and the biceps, triceps, supinator, knee and ankle jerks were all increased on the left compared to the right. No clonus could be elicited though stretch responses were very marked in all the muscles on the left. The left Hoffmann was positive, the right negative. The left plantar response was extensor, the right flexor.

The gait was abnormal. He dipped to the left when walking and the left foot was placed flat on the ground, more like an artificial leg than one of flesh and blood. When he walked or used the right hand the left tended to take up a position of marked adduction at the shoulder, flexion at the elbow, wrist and fingers and pronation in the forearm. When he turned the head to the right the left limb tended to take up a position of extension at the elbow though the other positions were unaltered.

Sensory findings were entirely normal. Superficial sensation was normally appreciated, when tested to light touch, pin prick, heat and cold. Joint and vibration senses were normal. Simultaneous bilateral pin prick showed better appreciation on the right. No Rombergism was evident, but when pushed he showed much greater tendency to fall to the right than to the left.

are the fine distal movements of the limbs affected, but the movements of the proximal joints, those designed to maintain the position of the limb steadily, also tend to be impaired.

As in dyskinesia, choreoid movements complicating hemiplegia tend to be exacerbated by stress, excitement and infections.

Lower motor neurone lesions.

Lower motor neurone lesions associated with hemiplegia have been largely ignored in recent years, though they were well recognised as being present in a few cases by earlier authors (Benedikt, 1874; Charcot, 1887; Briassaud, 1880). The latter attributed them to fibrous nodules in the degenerate long tracts of the spinal cord. With the increasing attention paid at the turn of the century to poliomyelitis, however, few cases showing lower motor neurone lesions were published as demonstrating the occurrence of mixed polioencephalitis and poliomyelitis (Strumpell's disease) (Lamy, 1894; Clark, 1912; Rothman, 1931). It is probable that most of these patients were in fact suffering from diffuse demyelinating encephalomyelitis (perivascular myelinoclasis).

In the present series muscle reactions were performed in six cases of hemiplegia, and in four of these, chosen because lower motor neurone lesions were suspected because of the presence of disproportionate muscle wasting in the affected limb, the presence of lower motor neurone lesions was confirmed. In two of the patients the lower motor neurone lesions were present in the peronei and anterior tibial groups. In the third wasting was apparent in the trapezius and deltoid, and /

Case 24.

Acquired hemiplegia with lower motor neurone lesions.

A boy of five at the time of examination who suffered from otitis media at the age of 9 months and after a severe convulsion showed left hemiplegia with muscle wasting in the left leg.

He was the first child of healthy parents, born in hospital after an uncomplicated pregnancy, labour and delivery, birth weight being 7 pounds 1 ounce. He thrived and developed normally until the age of 9 months at which time he was able to crawl and pull himself into the standing position. Ten days after the onset of fever and head rubbing which was diagnosed as being due to otitis media bilaterally he had a severe generalised convulsion with loss of consciousness lasting 90 minutes. He was admitted to hospital where the presence of fever, bilateral otitis media associated with cervical adenopathy was confirmed. No neurological abnormalities were noted. A lumbar puncture gave normal findings. His mother visited him next day and drew the attention of the staff to the fact that he was not moving his left limbs. On discharge ten days later he seemed drowsy and ill. He only gradually became as active as before during the next four weeks, and tended to neglect the left arm which was held across his chest, and to drag his left leg when crawling. He walked on his left toes when he began to walk without support at the age of 15 months. His left limbs became progressively dwarfed. His speech developed normally. He went to normal school at the age of five years, at which time he was able to play freely with other children but tended to neglect his left limbs, and was unable to use the left hand independently except for crude movements.

On examination. He was an intelligent small boy with marked wasting of the left limbs, affecting the lower limb more severely than the upper. The left upper limb was one third of an inch shorter than the right and the left lower limb a quarter of an inch shorter than the right. The circumference of the left thigh was one third of an inch less and that of the left calf half an inch less than those on the right. There was marked wasting of all muscle groups below the left knee, most marked in the anterior tibial and peroneal muscle groups. There was a severe talipes equinovarus deformity of the left foot.

There was a slight convergent strabismus. There was facial paresis of upper motor neurone type on the left. Power, co-ordination, reflexes and tone of the right limbs were within normal limits.

There /

There was impaired power in all voluntary movements of the left limbs, most severe below the knee. Fine and rapid movements were poorly performed by both arm and leg, and attempts to manipulate with the left fingers were very poor and accompanied by some athetosis. There was an increase of tone of spastic type in both left upper and lower limbs accompanied by exaggerated biceps, triceps, supinator, knee and ankle jerks and an extensor plantar response. He walked dipping to the right and showed a marked foot drop which made running impossible.

No abnormalities of sensation were noted. The left foot and hand were colder than those on the right.

Electrical reactions were performed using the Ritchie Sneath apparatus, and showed that increased current was required to produce contraction in the left tibialis anterior and peroneal muscle groups, whereas all other muscle groups required less than normal. These findings were felt to confirm the clinical impression that there was muscle atrophy in these groups.

and in the other in the supinator and extensor muscles of the forearm. All the cases were classified as suffering from severe hemiplegia. All four patients suffered from acquired hemiplegia and two were thought to be the result of acute disseminated encephalomyelitis, though in the other two the diagnoses were respectively cerebral thrombophlebitis and toxic encephalopathy complicating pneumonia. In these it seems possible that fibrous nodules in the degenerate long tracts may have encouraged on the anterior horn cells as shown so beautifully by Brissaud (1880). Case 24 is an example of a lower motor neurone lesion in hemiplegia.

It is hoped to perform electrical muscle reactions in a very much larger number of cases of hemiplegia in the near future. Unfortunately it is impossible at present to say how frequently lower motor neurone lesions occur in hemiplegic paralysis in childhood at present.

The importance of lower motor neurone lesions from the practical point of view is that they give rise to a form of contracture whose successful treatment will not result in any improvement of voluntary power in the affected muscles.

According to early pathological work the origin of the lower motor neurone lesions lies in the spinal cord where small nodules of fibrosis spread from the degenerate lateral tracts to involve the cells of the anterior horns (Brissaud, 1880).

Sensory impairment.

The classical authors found that sensory impairment occurred only occasionally in the involved limbs in hemiplegia (Lovett, /

(Lovett, 1888; Osler, 1889; Freud and Rie, 1891). Cases in which sensory impairment was prominent were described by Charcot (1887) however. In recent years the importance of sensory impairment has been increasingly realised (Tizard et al, 1954). The difficulties in assessing the frequency of sensory impairment are complicated in the younger and the more mentally defective patients by their inability to co-operate in testing, though as stated by Crothers it is generally possible to test a child of five fairly adequately if he is of average intelligence. Impairment of stereognosis was found by Tizard and his colleagues in approximately half of 106 patients with hemiplegia who were examined in detail.

Unfortunately detailed sensory testing was not as thoroughly performed as it should have been in the earlier patients in this series, though since the survey experience has abundantly confirmed the Boston findings.

Sensation to pin prick was found to be impaired in 10 cases, all of whom suffered from moderately severe or severe hemiplegia. Astereognosis was found in all of these, and impairment of stereognosis was observed in another 5, one of whom was classified as suffering from mild hemiplegia and 4 from moderately severe or severe hemiplegia, who did not show impairment of pin prick sensation. In all 15 patients there appeared to be some impairment of position sense.

Position sense was thought to be defective in the majority of cases of severe and moderately severe hemiplegia, but only in 23 was it thought to be severely enough affected to impair function more considerably than would otherwise have been the case. /

case.

Cerebellar ataxia was shown by four patients with hemiplegia. Three of these were classified as having severe or moderately severe hemiplegia and one as mild. The ataxia was manifest as past pointing and by deviation when walking either forwards or backwards blindfold. The ataxia was to the affected side in three of the four cases and to the other side in the fourth. Milder degrees of ataxia are probably fairly common in hemiplegia. It is extremely interesting to notice the variations in the ability to regain balance when the stance or position of hemiplegic patients in space is suddenly altered.

Neglect of the affected limbs.

Neglect of the affected side of the body in acute acquired hemiplegia has been recognised for many years (Jackson, 1868). But the frequency with which neglect of the limbs persists in these patients and with which it is found in congenital hemiplegia in childhood has not been generally recognised.

Neglect of the affected upper limb appeared to be more marked than neglect of the affected lower limb, and it was present in a marked degree in 42 of the patients, including all those noted to have impairment of stereognosis. Unfortunately, in the absence of the detailed sensory testing which should have been performed, it is impossible to relate the degree of neglect to the severity of the sensory deficits. It is only small comfort to find oneself in very good company in this respect (Crothers, 1954).

In general, neglect of the limbs tended to parallel the severity of the paresis, but not uniformly so, and in some cases perpetually /

perpetually reminding the child to use the affected arm appeared to be effective in improving function. Some mothers became remarkably adept at chanting, "Now use the right hand", "No, with the other hand" many hundreds of times a day.

Dwarfing in hemiplegia.

In contrast to diplegic paralysis, generalised dwarfing is not a feature of hemiplegia in childhood. On the other hand localised dwarfing of the limbs of the affected side has been noted from the time of the first studies of paralysis in childhood (Morgagni, 1762; Cazauvielh, 1827).

In this series of 75 cases only 7 showed no apparent shortening of the limbs. Measurements were not obtained in 4 cases, in 3 because of overactivity of the children. The remaining 61 cases all showed dwarfing of the affected limbs to some degree. Measurements were taken in the arm from the tip of the acronium to the tip of the middle finger, and in the lower limb from the anterior superior iliac spine to the internal malleolus. The degree of dwarfing found in the upper limbs of hemiplegic patients is shown in Table 59.

It is possible to compare easily the degree of shortening only in a rather rough and ready manner owing to the difficulties in allowing for the various differential factors of growth at different ages and in the different sexes in the three groups of hemiplegic patients. Analysis in detail would be an excessively complicated matter. However, the differences in age and sex distribution between the slight, moderately severe and severe cases are relatively small, and it is clearly not possible to account for the much higher proportion of severely dwarfed /

TABLE 59.

The apparent shortening in the affected arm, from acromion to top of middle finger in 75 cases of hemiplegia in childhood

Severity of the hemiplegia	Average age Yrs.	Mths.	No. of cases	No shortening	Less than 5"	More than 1.5", less than 1".	More than 1", but less than 1.5".	More than 1.5" but less than 2".	2" and over	Unknown
Slight	8	9	27	7	6	8	3	1	0	2
Moderately severe.	7	9	26	0	6	9	7	3	0	1
Severe	9	6	22	0	2	4	4	7	4	1
Total	8	3	75	7	14	21	14	11	4	4

dwarfed limbs in the severely paretic group on this basis. Though it is apparent that, in general, the severity of dwarfing tends to be in proportion to the severity of paresis, this is not invariably true. Thus 4 cases of mild hemiplegia showed more than 1 inch of apparent shortening in the affected arm, and 6 cases of moderately severe paresis showed dwarfing of less than 0.5" compared to the unaffected arm.

Similarly, though in general the severity of the dwarfing of the lower limb ran parallet to the severity of the dwarfing in the upper limb, this was also subject to variation. In most cases the apparent shortening from the anterior superior iliac spine to the internal malleolus in the affected compared to the unaffected leg was half that found in the upper limbs. Some cases showed almost equal shortening in the upper limbs and the lower limb, and in others the difference between the sound and the unsound upper and lower limbs was very marked. In three cases of severe upper limb dwarfing the difference between the lengths of the upper limb was more than four times that found in the lower.

The reduction of the girths of limbs was usually proportional to the apparent reduction in length, even when lower motor neurone lesions were present. In some cases where tendon lengthenings had been performed or calipers had been applied the reduction of girth in the lower leg was relatively more severe than the apparent reduction in length.

In general it was difficult to assess to what degree dwarfing of an upper limb impaired function, as severe dwarfing was usually associated with severe or moderately severe /

severe paresis, but in the leg it was of considerable importance. Though some compensation of reduction of lower limb length was achieved in many cases by the foot taking up an equinus position and by postural scoliosis, walking was still abnormal when this was present. Not only does shortening of one lower limb result in an unsteady and clumsy gait but also in scoliosis of the spine and a thoroughly unnatural posture being assumed to counteract its effects.

What became increasingly apparent as the survey progressed was how much more acutely conscious of dwarfing of the limbs some patients were than of actual paresis. This was especially marked in adolescent girls whose attempts at camouflaging the "baby arm" were often elaborate and always pathetic. One remarked that the only thing she wished for was for her arm to become normal for one evening so that she could go dancing in a strapless evening dress. Such extreme sensitiveness about the abnormal limb obviously tended to make the patient use it to a less extent than was actually possible, and favoured the spread of contracture.

Vasomotor disturbances in the limbs in hemiplegia.

In this survey temperature changes were not measured except by the back of the hand in comparison to the normal limb. Therefore the conclusions are in the nature of impressions rather than the result of scientific observation.

In the acute stage of acquired hemiplegia it was frequently found that the affected limb was warmer than the opposite side, the skin was congested, the pulse was fuller, and sometimes oedema of the foot and hand was noted.

In /

In one case the congestion of the limb and the swelling of the hand and foot was sufficient to suggest a diagnosis of acute lymphoedemato to the attendant surgeon.

After a few days, in most cases, the congestion of the limbs disappears, the oedema is absorbed and the limb becomes less warm. At the same time it begins to show excessive and somewhat erratic responses to changes in temperature, very readily becoming much warmer or much colder than the normal side. After a period of some weeks or months in the majority of cases the instability of temperature control is succeeded by a stage during which the limb is nearly always colder than the normal limb but in which it may be made very much warmer than the normal side by immersing some part of the body in warm water.

Some cases seem to recover from the initially vasomotor disturbance of their acute hemiplegia completely and temperature regulation seems to be grossly normal. A few cases show the permanently high temperature of the limbs noted so frequently in the acute stage, however. Two such cases were observed during the series and both were symptomless. Examples of cases showing the exaggerated reaction to heat as well as cold were encountered quite frequently but were classified with those showing a reaction to cold, as cold was the climatic condition which impaired the limb's function.

In most cases these stages are ill defined and they merge into one another, but at the same time the alterations in the type of vasomotor reaction may easily be observed in individual cases. In the majority the end result is that the affected limb /

TABLE 60.

Vaso-motor changes in the limbs of 75 children with hemiplegia

Severity of hemiplegia	No. of cases	Affected limb colder. Degree of difference			Affected limb always warmer	No difference in limbs.	Unknown
		Slight	Moderate	Marked			
Slight	27	12	2	0	0	13	0
Moderately severe.	26	16	3	1	1	3	2
Severe	22	10	5	5	1	1	0

Case 142.

A girl with acquired hemiplegia showing severe trophic abnormalities in the affected limbs following convulsions at the age of 7 months.

She was the second of four children born to healthy right handed parents after an uncomplicated pregnancy and a brief labour in hospital. Birth weight was 6 lbs. 8½ ozs. The child progressed normally until the age of 7 months at which time she became febrile, "off colour" and less aware of her environment than normal. Two days after the onset of symptoms she had a severe right sided convulsion with further impairment of consciousness which lasted for ten minutes. She was admitted to hospital. Occasional twitching of her right limbs continued after admission and she was observed to be very drowsy. Fever continued. Papilloedema was noted bilaterally. Lumbar puncture gave normal findings. The following day immobility of the right limbs was recorded. Treatment with penicillin was begun, and over the course of the next seven days her fever and papilloedema resolved and some movement returned to the right limbs.

She was finally discharged from hospital after six weeks, no definitive diagnosis of her original illness having been reached. She was restless, continuously active and showed a marked right hemiplegia. She stood with support at 18 months, and walked dragging the right leg at 22 months. Her first words were spoken at 22 months, but she was slow to say more than the simplest sentences. She began to use the right hand as an assistant to the left at the age of 2½ years and reached for objects with it independently at the age of 4.

During the second winter after her illness the right hand and arm were noted to go blue and sometimes black in cold weather. At times they were very swollen and she complained of pain in the hand. These complaints persisted every winter until the time of examination, when she was four years old.

On examination. She was an intelligent small girl with some difficulty in expressing herself and some retardation of speech development. The left cranial hemicircumference was three-quarters of an inch less than the right. The right upper limb was one and seven-eighths inches shorter and the lower limb seven-eighths of an inch shorter than the left.

There was a moderately severe right facial paresis of upper motor neurone type. Voluntary movements of the right upper limb were very limited. Only a few degrees of supination were possible and the fingers could not be fully extended nor the thumb abducted. She made good use of such movements /

movements as she had, however, and could pick up and handle objects with the hand. There was limitation of dorsiflexion of the right foot and she could not move the right toes. Proximal movements of the limbs were less affected than distal movements.

There was a marked spastic increase of tone in the right limbs compared to the normal tone on the right, and the right biceps, triceps, supinator, knee and ankle jerks were markedly increased. The right plantar response was extensor, the left flexor. The right Hoffmann response was present. When she walked she dipped severely to the right and walked on the right toes, dragging the right leg to some extent. Marked associated flexion movements of the right upper limb were evident. No abnormalities of sensation were found. The right hand and foot were bluish in colour and appeared slightly swollen. They were much colder to the touch than those on the left, and pitting oedema was present on the dorsum of the right hand.

limb is colder than the normal limb in ordinary temperatures. It tends to become very much colder on exposure to temperatures below the usual, and may become cyanosed and swollen in even mildly cold weather. There may or may not be over reaction in the limb to warm conditions.

This is the most commonly encountered type of disturbance in the hemiplegic limbs, and for purposes of classification they have been divided into three categories, mild, moderate and severe. Mild cases are those in which cyanosis is sometimes evident. Severe cases are those in which cyanosis tends to be persistent or very readily produced and in which oedema someyimes results from cold. Case 142 is an example of the latter type.

Of the 75 cases of hemiplegia in the series, 16 showed no gross disturbance of temperature regulation in the affected hand. Thirty-nine cases showed slight disturbance, 10 showed moderately severe disturbance, and 6 showed severe disturbance. Two cases showed an increase of temperature in the affected arm, compared to the normal which was permanent and apparently no handicap. (Table 58.)

Of the cases classed as being severe, all except one were also classed as cases of severe paralysis; of those classed as suffering from moderately severe vaso-motor disturbance, two were classed as mild cases of paresis, 3 as suffering from moderately severe paresis and 5 as suffering from severe paresis. It will be observed that the figures in Table 58. indicate that, though in general the incidence and severity of vasomotor disorder bears some relationship to the severity of the paresis, this /

this is by no means always the case. One case of severe paresis showed no apparent difference in the temperature of the limbs and in another the affected hand was permanently warmer than the other.

The importance of the vasomotor disturbance in the limbs in children with hemiplegia appears to be somewhat underestimated. In moderately severe and severe cases the affected limbs may readily become so cold that their function is severely impaired during cold weather. The contrast in the performance of a child in a slightly cold room with one in a normal or slightly warm one is striking. It was no accident that one child, made to use the affected hand at school, remarked that he got good marks for writing in summer but bad marks in the winter.

In some of the more severe cases pain as well as loss of function results from the exaggerated response to cold in the damaged limbs.

Contracture.

In the early stages of acute acquired hemiplegia absence of some degree of voluntary movement may be demonstrated in the limbs in most cases, though these movements may be full when the limb is handled passively (Twitchell, 1951).

Unless these movements are frequently actively attempted or accomplished passively a change in the state of the limbs occurs. Little used or neglected movements become impossible either actively or passively because of contracture, commonly appearing from 3 to 6 weeks after the appearance of hemiplegia in acquired cases. Because there is a tendency to neglect to utilise impaired voluntary movements to the full, restriction of /

of movement due to contracture usually overshadows the restriction of movement due to loss of voluntary power.

Contracture is therefore most likely to be extensive and severe in those with severe loss of voluntary power, who cannot move their limbs, in mentally defective or very young children who cannot be made to use their limbs, or in those in whom no attempt is made to keep the limb mobile.

The the liability of contracture to affect little used limbs is not fully appreciated is indicated by the numbers of mentally defective patients encountered in institutions, whose neurological disorder is hemiplegic but whose contractures are generalised. Because of the paralysis of one side the child finds movement difficult. Because he lacks desire to move he is allowed to reach the state where he cannot move because of contracture of the sound as well as the damaged side.

In cases showing severe contracture it is impossible to gauge to what degree apparent inability to move an affected limb is a true reflection of the extent of the underlying loss of movement patterns.

The position of limbs showing contracture was very constant. The limbs took up positions of flexion because the extensor muscles tended to be more affected by paresis than the flexor muscles. The distal parts of the limbs were more affected than the proximal, and more primitive movements were less affected by contracture than those of later development. Thus the thumb tended to be flexed and adducted into the palm, the fingers flexed, the wrist and elbow flexed, the forearm pronated and the shoulder adducted.

OTHER FINDINGS IN HEMIPLEGIA.

Under this heading will be considered the other abnormalities encountered during the examination of the 75 cases of hemiplegia in the survey.

Intellectual impairment.

The assessment of intellectual ability in cases of cerebral palsy is a matter of the greatest difficulty. In the first place the child may be incapable of showing his intelligence because of motor impairment. In the second place his intelligence may not have had a chance to develop fully because of the restricted experience and limited environment he has lived in. Not only may his environment and experiences be limited by his motor handicap but the senses may be directly affected; Some are aphasic and have specific difficulties in the comprehension and expression of their ideas. Obviously such handicaps cramp the development of intellectual abilities.

On intelligence testing, therefore, performance may not be a true measure of the child's actual intelligence, because of his motor handicap. Even when it is a true measure of his actual intelligence it is no estimate of the intelligence which he may be capable of developing in an ideal environment. Thus intelligence tests can only be regarded as very rough guides to actual or potential intellectual capacity.

A patient's social and educational achievements are also poor measures of his intellectual capacity in cerebral palsy, for many severely paralysed cases with normal intelligence show marked retardation in their developmental milestones and

in /

TABLE 61.

The intelligence and schooling of 75 cases of hemiplegia in childhood

Intelligence Quotient	115+	100-114	85-99	70-84	55-69	Less than 55	Untested
Severity of the paresis. (all patients)	3	10	10	17	17	11	7
Mild	1	2	4	4	8	5	3
Moderately severe	2	4	2	4	7	4	3
Severe	0	4	4	9	2	2	1
Normal schools	1	3	9	8	2	0	0
Schools for the physically handicapped.	0	1	2	4	3	0	1
Schools for the mentally handicapped.	0	0	0	6	8	1	0
Ineducable	0	0	0	0	1	6	2
Not yet at school	0	2	1	2	1	4	7

in their educational and social progress.

Yet, although they are fallacious, the only useful measures of the child's intellectual ability that are available are the levels of his performance on intelligence testing and in social and educational achievement. Experienced observers are able to assess intellectual capacity on the basis of these findings in a large proportion of patients, though a long period of observation under standard conditions may be necessary (Dunsdon, 1952).

In Table 61 are shown the intelligence quotients found in a number of patients with hemiplegia by different psychologists working under different conditions and using different tests. In addition the levels of educational achievement are roughly indicated by noting the educational placement of the child in normal school, special school or institution.

It will be observed that 28 of the 68 tested patients (approximately 41%) had I.Q.s beow 70, and only 23 (33%) above 85. These figures give a distribution of hemiplegic patients by I.Q. comparable to that observed by Dunsdon (1952) and Hodd and Perlstein (1955). Thus, though the testing in this series of patients was far from systematic it is probable that the final results are not too wide of the mark. It will be noted that there appears to be little relationship to severity of physical handicap.

Unfortunately it was not possible to investigate the patients systematically using similar tests in all during the survey, as was really desirable. The Wechsler test, in particular, with its separate performance and verbal scales

is /

is capable of giving information of great value, not only about the intellectual endowment but about the nature of the loss of specific abilities so commonly found amongst hemiplegic patients.

Though the educational difficulties of all patients with cerebral palsy will be discussed later it is worth emphasising here that specific impairment in the ability to deal with written symbols, an inability to organise and retain in the memory the organisation of masses in space and confusion of the horizontal, vertical and third dimension were very commonly found in hemiplegic patients. These difficulties tend to reduce the scores in performance testing below those in verbal testing, whilst verbal scores are usually lower in aphasic patients than are the performance. These forms of mental impairment suffered by hemiplegic patients are remarkably similar to those found in patients with so-called developmental aphasia or dyslexia (Ingram and Reid, 1955). They have been studied in detail by Hood and Perlstein (1955) and Dunsdon (1952). Probably the best description of them is in a series of cases following measles encephalitis, however, though only a minority of these patients had residual hemiplegia (Byers and Meyer, 1951).

The level of intelligence varied from the apparently normal to the grossly subnormal and untestable. But that I.Q. alone was not always a good guide of the level of functioning is sufficiently indicated by the wide spread of scores amongst patients in similar types of school.

In patients suffering from hemiplegia of congenital type and /

Case 117.

Girl with acquired hemiplegia illustrating the disparity between motor and speech milestones which may be present in mentally retarded patients.

The only child of healthy parents married eight years before her birth, at which time mother was 37 years of age. Subsequent abortion ten years later.

Delivered spontaneously by the vertex at home after uncomplicated pregnancy and an abnormally brief labour of one hour. Immediately after birth the child seemed normal and cried. Birth weight was 7 lb. Artificially fed and thrived for two weeks and seemed normal. Then began to vomit in projectile manner and lost weight. Became drowsy and lethargic, progressively difficult to feed. From age of three weeks she showed generalised epileptic seizures of brief duration five or six times a day. Admitted to hospital aged five weeks weighing 4½ lb. After resuscitation and correction of extreme dehydration Rammstedt operation performed for congenital pyloric stenosis. Thereafter thrived, but brief generalised seizures continued to occur. On account of these she was seen periodically in the out-patient department. At the age of four months it was noted that the right hand was not used for holding offered objects though the left was and that there was spasticity and restriction of supination in the right forearm. By this time she was sitting with support, and she was walking without support shortly before the age of one year, though it was observed that the right heel was not placed to the ground when she did so. She used her right hand only at the age of four years as an assistant to the left.

Her speech was very slow in developing; her first intelligible words, apart from "Mum" and "Dad", were said at the age of three and a half years, and she only had phrases at the age of eight or nine years. She had never been to school and attended an occupation centre where she was found to be toilet trained and "no trouble". Her fits ceased at the age of about four years. On examination at the age of fourteen she was found to be an ungainly adolescent girl who was clearly mentally retarded.

There was a moderately severe spastic right hemiplegia with 1" dwarfing in length of the right upper limb compared to the left and ½" reduction in length of the right leg compared to the left. Sensory findings were probably normal.

She was able to answer simple questions but her vocabulary was small and her syntax and pronunciation grossly immature. There were multiple defects of articulated consonants. She could count only to five correctly, and though she knew her name she did not know her age, birthday or address.

Case 191.

Slight right hemiplegia following measles encephalitis, illustrating good recovery.

The only child of healthy parents, both right handed. Mother was 20 at the time of delivery. She was well in pregnancy and was delivered in hospital at term after a labour of 7 hours, spontaneously under chloroform analgesia. The birth weight was 9 lbs. 14 oz. His development and motor and speech milestones were within normal limits. He was rather a quiet boy.

At the age of 5 years he developed measles, and six days after the development of his rash, when he had seemed to be improving, he became very drowsy, lost his appetite and complained of headache. The doctor found that he had neck retraction and that he was very fevered. He was admitted to hospital where a diagnosis of measles encephalitis was made. He appeared to be confused, unable to obey commands, and not fully oriented for the first week of his stay. The following week he appeared to be unable to see, was stuporose and quite unresponsive to his parents. He had one slight generalised convulsion with loss of consciousness. Lumbar puncture findings were normal. Thereafter he improved and was discharged after nine weeks in hospital.

On discharge his whole nature was changed. He was demanding and could brook no frustration. His temper was labile and variable. He was overactive and curious, and never still for a moment. His speech was badly pronounced and most of it was impossible to understand because he ran words into one another. He made no effort to use the right hand which was "held cramped up against the chest". He limped and dragged the right foot. His vision was very poor.

In the year following his discharge from hospital his overactivity became less, his speech improved so that only when he was excited was it then incomprehensible, and his walking became steadier. After six months he began to use the right arm to assist the left and after one year was able to use the right hand independently, though it was still clumsy. After 18 months he was able to do buttons with the left hand assisted by the right. His vision appeared to be normal. He attended normal school after 6 months at home and has done quite well there, being in the middle of his class with children of his own age.

On examination. Height 45½ inches. Head circumference 21". The right arm was one-eighth of an inch shorter than the left. The legs were of equal length.

He /

He was alert and quick in response to commands or questions, but not always accurate. He tended to be facile rather than clever. He showed flights of ideas related only by chance word associations. His speech was normal and his comprehension was good. There was a right facial paresis of upper motor neurone type, moderate in degree. The power and co-ordination of the right arm and leg were impaired. The grasp was moderately good and large objects were firmly held. On the other hand small, fine or rapid finger movements were very poorly performed. All movements of the right hand were accompanied by marked athetosis of the ring and middle fingers which extended and separated. The athetosis was much less in the index and little fingers and was slight in the wrist, so that the athetosis by itself did not grossly impair his efficiency.

The only movement that was limited in extent on voluntary effort was supination of the forearm, but abduction of the shoulder and extension of the elbow, wrist and fingers were weaker than on the left. In the leg dorsiflexion of the foot and extension of the knee were weaker than on the left.

The gait was abnormal with the foot being in a position of slight equinus and the knee slightly flexed. When he walked or used the left arm the right arm tended to assume a position of adduction at the shoulder, flexion at the elbow, wrist and fingers, and pronation of the forearm.

There was moderate increase of tone of spastic type in the right limbs and the biceps, triceps, supinator, knee and ankle jerks were increased on the right compared to the left. The right Hoffmann response was present, the left absent. The right plantar response was extensor, the left was flexor. Sensory findings were similar and normal on the two sides. The right hand and foot were considerably colder than the left.

and in those in whom it was acquired shortly after birth, who were severely mentally defective, milestones were usually somewhat retarded, but not consistently so. Whereas speech development was usually very slow motor milestones were often much less affected. Case 117 is an example of such a patient.

In some patients, especially those with frequent epilepsy, there appeared to be some deterioration of performance on intelligence testing, and some patients appeared to stick at a given mental age when they were 10 or 12 years of age. Unfortunately the data is not sufficiently extensive for an attempt to be made to see if patients with epilepsy have a significantly lower I.Q. than those without fits, as reported by Hood and Perlstein (1955).

The length of time for which patients tended to improve in performance on intelligence testing and in learning after acquired hemiplegia was often remarkable, as has also been noted by Byers and Meyer (1951). Case 191 is an example of the very great recovery which can be attained over a prolonged period.

Aphasia.

Interference with the reception and expression of ideas in speech was found relatively commonly in cases of hemiplegia. In those with mental defect of moderate or severe degree it was almost invariable, but overshadowed by the intellectual impairment. Cases in which the interference with the reception or expression of ideas was clinically recognisable, either because it was a relatively isolated or severe disability, were classified as aphasic. There were 12 such cases.

In /

In view of the controversy, especially rampant in the late years of last century as to whether aphasia was at all dependent on the side of the brain damage, it is interesting to note that all but one of the cases of aphasia showed right hemiplegia. The exception was left handed before the onset of his acquired hemiplegia (Bernhardt, 1885; Marie, 1888; Dreyfous, 1882).

Though the numbers are too small for any statistical treatment it is interesting to note that 8 of the 12 cases occurred in cases of acquired hemiplegia. Many cases from whose histories it seemed likely that aphasia had been present in the acute stages of the acquired hemiplegia showed no such abnormality at the time of examination.

In some cases the degree of aphasia was severe, and ability to imitate speech in these children was very striking compared to their inability to express their ideas. In milder cases the aphasia was more of a nuisance to the child because it resulted in frequent word misplacements and substitutions rather than a very serious disability. It is worth emphasising that abnormalities of speech, particularly sound distortion, substitutions and alterations of order of syllables and rhythm are almost always found in aphasic patients.

In Table 62. are shown the numbers of cases of aphasia which occurred in patients showing mild, moderately severe and severe paresis. It will be noted that aphasia occurs relatively less frequently in cases of mild hemiplegia. Case 192 showed severe aphasic speech disorder.

Articulatory /

TABLE 62.

Aphasia and speech difficulties in 75 cases of hemiplegia in childhood.

Hemiplegia	No. of cases	Aphasia	Articulatory defects	Stammer or hesitation	No comprehensible speech	All types of speech defects
Right	54	11	8	3	2	13
Left	21	1	6	1	1	8
Slight	27	2	7	2	2	11
Moderately severe	26	6	6	1	1	8
Severe	22	4	1	1	0	2
Normal schools	23	1	0	0	0	0
Special schools	26	4	10	2	0	12
Ineducable	9	3	1	0	2	3
Under 5	17	4	3	2	1	6

Articulatory defects.

For the purposes of the survey only those whose speech was so severely defective that comprehension of it was a matter of difficulty were included. Cases whose vocabulary was poor but pronunciation normal or adequate to make their meaning clear were not classified as having speech defects.

Three cases of hemiplegia, all severe in type and two of them right-handed, had no speech or only a few very ill-pronounced words. Fourteen suffered from inability to form sounds correctly which was severe enough to make them largely incomprehensible, and 4 cases stammered. One of the latter was a case of congenital hemiplegia who began to stammer at the age of 6 years, the others had begun to stammer shortly after beginning to say sentences.

Of the patients who were unable to say sounds comprehensibly, only 3 appeared to be dysarthric. In the others the defects were developmental in type, being similar to those encountered in patients with developmental aphasia or "articulatory apraxia" (Morley, 1955). They were commoner in the more mentally retarded patients.

This relatively small incidence of speech defects compared to other series is, of course, the result of the definition of what constituted speech defects. If even minor deviations from normal pronunciation and articulation are considered, then a very high proportion of patients had abnormal speech, as reported by Dunsdon (1952). But in the majority of such patients the speech defects, though interesting academically, cannot be considered to be a significant disability.

In /

In Table 62. are shown the numbers of cases with speech defect attending normal and special schools and considered ineducable. It is interesting to note that no child attending normal school had speech severely enough affected to make it difficult to comprehend. This may partly be due to the fact that such children, whatever their intelligence, tend to be transferred to special schools on account of their defective speech.

Visual defects.

Visual defects of various types are common in hemiplegia, and were well recognised by earlier German authors and discussed in much detail by them (Koenig, 1895; Freud, 1889).

The common defects in this series were of two types, errors of refraction and amaurotic. Five cases of severe myopia were recorded. Four of these cases were mentally defective.

Of much more clinical importance are the amaurotic disorders. The types and numbers of these encountered are shown in Table 63.. It will be seen that severe and moderately severe cases of hemiplegia show amaurotic defects more frequently than do slight cases. Two cases of hemiplegia showed no useful vision. Both these cases were of congenital origin and in both bilateral optic atrophy had occurred.

Two of the cases showing unilateral amaurosis were of great interest. Both were amaurotic in the eye opposite to the side of the paresis. In both the retinae showed extremely scanty vessels and very white optic discs with irregular margins. The appearance was of severe unilateral optic /

TABLE 63.

Amaurosis in 75 cases of hemiplegia in childhood

Type of paresis	No. of cases	Complete amaurosis	Unilateral amaurosis	Field defect	Totals
Right hemiplegia	54	1	1	5	7
Left hemiplegia	21	1	2	1	4
Severity					
Slight	27	1	0	0	1
Moderately severe	26	0	1	3	4
Severe	22	1	2	3	6
Totals	75	2	3 *	6	11

* Two cases of unilateral optic atrophy are of postnatal injury.

optic atrophy probably the result of retinal thrombosis. In both carotid pulsation in the fauces was equal. Possibly the involvement of the retinal vessels was related to involvement of the cerebral vessels which could be held responsible for the hemiplegia - a speculation too difficult to resist.

The cases showing field defects did so consistently to the side of the paresis. The degree of field defect varied greatly in extent. In one case epileptic fits commenced with phenomena appearing in the defective quadrant.

Involvement of other cranial nerves.

The eyes. Slight asymmetry of the pupils and reactions to light and accommodation being somewhat brisker on one side than the other were encountered not infrequently in cases of hemiplegia, without apparent visual defect or other evidence of oculometer involvement. This finding is in accordance with the old observations of Koenig (1896).

Strabismus occurred relatively commonly in cases of hemiplegia in childhood, and limitation of eye movements was noted in 22 cases. In 13 of these one eye was affected, in 9 both eyes. In three cases the oculometer nerve was affected, in 19 cases abduction of the eyes was limited. No deductions were possible about the aetiology of the strabismus. It was stated to have been present in a number of other patients during infancy but had gradually become less marked and was absent at the time of the examination. Most of the cases of strabismus tended to show improvement during childhood, and the majority in which abduction of the eyes was limited appeared to be of neuromuscular rather than central origin.

There /

There was no greater incidence of strabismus in the eye on the affected, than on the unaffected side.

Facial palsy. Facial palsy was stated to be uncommon in hemiplegia in childhood in most of the earlier reviews (Osler, 1889; Lovett, 1888). In this and other recent series, however, it has been found to be present, at least in latent form, in the majority of cases.

The form of the paralysis varies somewhat in type and greatly in severity in different cases, as noted by Koenig (1896) and Freud (1897).

The various forms encountered are indicated in Table 64. Though the facial paresis tended to be more apparent in cases with moderately severe or severe palsy of the limbs, a number of mild cases showed quite marked facial involvement.

It will be seen that 14 cases showed no abnormality of facial movements. Twenty-one cases showed asymmetry of the face at rest and on movement, and 31 appeared to show facial symmetry at rest, but on movement, whether emotional or voluntary in type, the affected side moved less than the normal and asymmetry was produced. A smaller proportion of cases showed very interesting types of abnormality. In 5 cases the face was symmetrical at rest, but on movement of voluntary type the affected side showed definite lag. On movement of emotional type, however, the affected side showed overaction, and the asymmetry appeared, literally, to be on the other cheek. Four cases showed symmetry at rest and on emotional movements. But on voluntary movements a lag was present. The types of facial paresis encountered are similar to /

to those described by Freud and Rie (1891) except that he found a larger proportion of cases who showed overaction on emotional movements. In his series approximately the same proportion of cases as in this series (70%) showed little or no asymmetry of the face at rest.

Hearing. A small number of cases showed middle ear deafness due to chronic otitis media with or without mastoiditis, which had indirectly led to the hemiplegia in a number of cases. One case whose pneumococcal meningitis had been treated with streptomycin showed severe, but not complete nerve deafness. Two of the cases found to be aphasic had been diagnosed previously as being deaf.

Swallowing. The frequent drooling seen in infancy in cases of hemiplegia appears to be due in most cases to infrequent swallowing. It is more common in mentally defective children than in others. It tends to improve at about the same age as the child learns to speak.

When the frequency of swallowing is noted in drooling children it is quite frequently found that they swallow as infrequently as once in two and a half or three minutes. Clearly this phenomenon may be due to the mental inertia of the child or to true difficulty in swallowing. In three cases, aged respectively 9, 3 and 3½, all with great speech difficulty and showing drooling, swallowing of solid food was difficult and the palatal reflexes were abnormal, sluggish in one case and asymmetrical in two. Only one of these children appeared to be severely mentally defective.

Though /

TABLE 64.

Strabismus in hemiplegia

Type of hemiplegia	No. of cases	Unilateral involvement		Bilateral involvement		Total
		Oculomotor	Abducent	Oculomotor	abducent	
Mild	27	0	3	0	1	4
Moderately severe	26	1	4	1	6	12
Severe	22	1	4	0	1	6
Totals	75	2	11	1	8	22

TABLE 64a

The facial paresis in 75 cases of hemiplegia

Types of facial paresis	Number of cases
No abnormality evident	14
Facial asymmetry at rest and on movement.	21
Face normal at rest but lagging on the affected side on all movements.	31
Facial asymmetry apparent only on voluntary movement, not on emotional movement, or at rest.	4
Face lags on voluntary movement but overacts on emotional movement. Symmetrical at rest.	5
Total	75

Though relatively few cases with true difficulty in swallowing were found it seems possible that a number of cases of congenital hemiplegia do in fact have definite neurological cause for their difficulty in swallowing, and therefore feeding, in their first few months of life.

Shoulder movements. Two cases were encountered in whom there was weakness of voluntary head rotation away from the side of the lesion. One of these cases showed some winging of the scapulae. One was classified as a moderately severe case of hemiplegia and showed facial, tongue and speech involvement. The other was a severe case without any other evidence of cranial nerve involvement but with definite dwarfing of the trunk on the affected side.

Tongue involvement. Involvement of the tongue was encountered much more frequently than expected and was present in 9 cases. In 3 involuntary movements of the tongue were evident when it was produced. In one case these were severe and appeared to be generalised. In the other two cases they were slight and confined to the side of the paresis. Only the case with generalised involuntary movement showed speech defect.

In 6 cases some weakness of the tongue was evident on voluntary movement, and the tongue tended to be deviated to the paretic side when central protrusion was attempted. Two of the 6 cases showed marked speech defect, but the speech of the other 4 was easily comprehensible. Of the cases showing paresis of the tongue, all were classified as severe or moderately /

moderately severe cases of hemiplegia.

Head asymmetry. The fronto-occipital circumference and hemicircumferences were measured in all those in whom it was possible to obtain reasonably accurate measurements. The skull was found to be asymmetrical by ½" or more on measurement in 9 cases though it was plagiocephalic in a much larger number. Only two patients with acquired hemiplegia showed a difference in the measurements of the affected side of the cranium and the unaffected, whereas 7 cases of congenital, or probably congenital, origin did so. The commonest asymmetry was a flattening of the patiental or patiental and occipital regions on the side of the head opposite to the paralysis. This was not always marked enough unfortunately to produce any significant difference on measurement, however striking to the eye.

Variations in the head circumference in hemiplegic and normal children, and the incidence of various head shapes will not be discussed as measurements of controls suitable for comparison are not yet available.

Trunk asymmetry. Unfortunately routine measurements to determine trunk symmetry were not made though the trunk was measured bilaterally in most cases showing marked shortening of the limbs. In only 3 cases was the chest hemicircumference found to be smaller by ½" or more on the paralysed side. Two of these cases were severe and one moderately severe cases of paresis. All showed marked limb shortening on the affected side.

Epilepsy /

TABLE 65

Epilepsy in 75 patients with hemiplegia in childhood.

Severity of hemiplegia	Number of cases	Type of attack			No. of patients with epilepsy (6 with more than one type of attack)
		Grand mal	Jacksonian	Myoclonic or Petit mal	
Mild	27	4	2	0	5
Moderately severe.	26	7	3	4	12
Severe	22	4	4	0	7
	75	15	9	4	24

Epilepsy in hemiplegia in childhood.

Broadly speaking the conclusions that Wuillamier (1882) reached about the occurrence and nature of epileptic phenomena in hemiplegic paralysis were accepted and have been found to be true for the majority of cases in this series. All but three of the acquired cases of hemiplegia showed convulsive attacks in the acute stage of the initial illness. A large proportion of congenital cases showed convulsive attacks in the first few days of life. The nature of the initial convulsions varied greatly. Some showed generalised clonic and tonic manifestations, but the majority showed the most marked manifestations on the side later found to be paralysed. In most cases the fit began in the hemiplegic side, and though in most cases after the preliminary convulsive movement the fit became generalised, clonic movements were usually more marked on that side throughout the fit. The severity and the duration of the initial fits varied greatly.

As the acute stage settled and the hemiplegia became apparent the fits ceased in the majority of cases, but they persisted in 5 cases without remission. In the remaining cases they were absent for variable periods of time but then convulsive attacks, not necessarily of similar type to those initially present, recurred.

Chronic epilepsy, the end result of the former series of events, was present in 24 cases of the 75 cases of hemiplegia in the series, as shown in Table 65 . It will be seen that 4 cases showed more than one type of epileptic attack. Three showed petit mal or myoclonic epilepsy and grand mal, and one

showed /

TABLE 67

Frequency of epileptic attacks in 24 children with

Type of epilepsy	No. of cases	More than one per day	More than one per week	Less than one per week. More than one per month.	Less than one per month. More than one per year.
Grand mal	15	0	4	3	8
Jacksonian attack.	9	0	7	1	1
Myoclonic jerks or Petit mal.	4	4	0	0	0
	24 *	4	11	4	9

* had more than one form of epileptic attack.

TABLE 66

Period of freedom from fits in 24 hemiplegic children with epilepsy

Type of epilepsy	No. of cases	No freedom	Less than one year	Over year but less than 2 yrs.	2 yrs. but less than 3 yrs.	3 yrs. or more
Grand mal	15	2	3	6	1	3
Jacksonian attack.	9	3	1	2	3	0
Myoclonic jerks or Petit mal.	4	0	4	0	0	0
	24 *	5	8	8	4	3

* 4 patients with attacks of more than one type

showed petit mal associated with Jacksonian attacks. The latent periods following the appearance of the hemiplegia during which no epileptic attacks occurred are shown in Table 66 . It will be seen that the interval was less than two years in the majority of cases, including all those in whom the only apileptic disturbance was petit mal.

Three main forms of epileptic attack occurred, though variations of these were not uncommon. The commonest was grand mal. Some of these cases showed the most marked clonic movements during the attacks on the side of the paralysis, but in all the attacks were generalised rather than localised and began suddenly, not being confined initially to one limb. The next most frequent form was Jacksonian in type, the first abnormal movements occurring in the affected limbs, and generally because consciousness was slowly lost or not lost the child could localise its origin precisely. Petit mal alone occurred in only 3 patients, and one had myoclonic jerks.

Two cases with Jacksonian attack were of especial interest, one showing predominantly sensory abnormalities in the initial stage of her attacks, and the other, previously quoted, experiencing visual hallucinations.

The severity of the attacks and the frequency of their occurrence varied greatly. In Jacksonian attacks loss of consciousness tended to be more gradual, less profound, and attacks were accompanied by more premonitory symptoms than were found in attacks of grand mal type. This was of obvious importance from the point of view of the child recognising his attacks and being able to put himself into conditions of relative safety before they were fully developed.

The /

The frequency of the attacks of each type is shown in Table 67 . That epileptic attacks appeared to be commoner in those with moderately severe hemiplegia than in those with slight or severe hemiplegia is shown in Table 65 . Possibly the severe cases had more destroyed and less damaged cerebral tissue than had the moderately severe cases.

One point of some clinical interest was that in some patients the hemiplegia was apparently more severe following severe epileptic attacks for as long as two weeks after these had ceased, a matter of obvious importance so far as limb function was concerned.

As shown in Table 68 , the majority of patients with epileptic attacks received anti-epileptic drug therapy. Most patients received several drugs alone or in combination at different times. They were successful in reducing the numbers of epileptic attacks in the majority.

Behaviour disorders in hemiplegia.

Functional behaviour disorders occurred commonly in patients with cerebral palsy of all types. They were most frequently due to the child feeling frustrated as a result of his physical and mental disabilities and to his inability on account of them to compete with other children. Others were unable to adapt to society because of being over-protected at home. This tended to be commoner in cases of congenital cerebral palsy in which families were smaller than in cases of acquired cerebral palsy. The extent to which unrecognised educational difficulties were responsible for children becoming frustrated and disheartened appeared to be great.

The /

TABLE 67

Effect of therapy on frequency of attacks in 24 hemiplegic patients with epilepsy.

Type of attack	No. of cases	Having therapy	Attacks less frequent on therapy.
Grand mal	15	12	10 *
Jacksonian	9	6	4
Myoclonic jerks or Petit mal.	4	2	2

* including 2 with no attacks for 2 years on therapy

The nature of the functional behaviour disturbances did not appear to be different in type from those in non-handicapped children. Temper tantrums, enuresis, masturbation, solitariness, night terrors, excessive fearfulness and petty delinquency were all encountered.

The most common behaviour disorder which did not appear to be of functional origin but to be dependent upon the presence of brain damage was a very characteristic type of restlessness and overactivity. This was associated with a very short attention span, inability to concentrate, a compulsive desire to touch everything and often put them to the mouth, fluctuations of mood, lack of affection and outbursts of aggressive and irresponsible behaviour. This form of behaviour was evident in 9 patients with hemiplegia and occurred in 2 congenital and 7 acquired cases. Six were thought to be ineducable, and 4 were epileptic.

The history of the development of the overactivity was rather characteristic in the acquired cases. Initially after the occurrence of cerebral damage the patients were noted to be quieter than usual, and some were definitely lethargic for a period varying between 6 weeks and 4 months. Thereafter lethargy was gradually replaced by increasing activity and interest in the surroundings, restlessness and a tendency to be "interested in everything at once rather than in any particular thing", as one mother put it. This interest was manifest as a tendency to touch everything within reach and attempt to reach any object which was not. The object would be touched, put to the mouth, sucked or chewed and then discarded /

Case 144.

Left hemiplegia, epilepsy, overactivity and mental deficiency following cerebral thrombophlebitis.

The second child born to healthy right handed parents after uncomplicated parturition, birth weight 7 pounds. The neonatal period was normal. At the age of 4 months the child developed a discharging right ear. In spite of treatment the ear continued to discharge for a matter of months. His motor and speech milestones were within normal limits.

At the age of 22 months he developed a head cold. The right ear discharge increased and after one week of fever, listlessness and anorexia associated with the catarrh he had a convulsion with sudden loss of consciousness and generalised twitching of all limbs. He was admitted to hospital and was noted to show generalised increase of muscle tone, fever, a discharging right ear, positive Babinski signs bilaterally, and normal lumbar puncture findings. During the first three days of his stay in hospital a left hemiplegic paralysis became defined and a right jugular thrombosis was noted. Fever persisted and he was transferred to a neurosurgical unit. He was treated with streptomycin and heparin and the fever finally settled five weeks after his convulsion, and he was discharged back to the original hospital, where he developed whooping cough followed by measles. He was eventually discharged home six months after his convulsion. At this time he was just beginning to stand again. He was miserable, lethargic and tended to sit or lie whining most of the day. He could say nothing and used the left hand hardly at all. The mother noted that there was a squint which had not been evident before his admission to hospital.

When he walked he tended to drag the left leg. His appetite was good and he soon showed a marked tendency to put anything he handled into his mouth and chew his clothes. Gradually over the course of a year a change took place in his behaviour. On discharge from hospital he had tended to be drowsy and slow. He slept a great deal and noted little, but as time passed he began to wander about more and more, handled everything in sight and put them into his mouth. He became progressively destructive, tearing and eating anything he could reach. He did not seem to be able to comprehend anything that was said to him, and attempts to control him seemed to make his destructive behaviour worse. Eighteen months after his discharge from hospital he did seem to begin to understand simple commands.

At the time of his return home from hospital he had fits every day or every other day. These started gradually by his going pale, the eyes glazed, and he would lose consciousness for about ten minutes during which time the left side only /

only showed clonic movements of the limbs. After recovery of consciousness he would sleep for several hours. Since his discharge from hospital the fits have gradually become less frequent and less prolonged, and in the past 18 months no more than five minutes are accompanied by a tonic state of the left limbs but no clonus, and occur only once a week or so. His behaviour has remained unaltered and he still has no speech. He uses the left arm only when he has to and only to assist the right.

On Examination. Height 41¼ inches. Head circumference 20½ inches. There is half-an-inch smaller measurement on the right than the left hemicircumference of the skull. There was one-third of an inch shortening in the left arm and one quarter of an inch shortening in the left leg compared to the right.

The child was grossly overactive in behaviour. He was never still. He fiddled with objects continually and chewed anything in sight. He paid no attention to or could not comprehend commands. He shrieked but had no speech, and could not imitate. There was a bilateral paresis of the sixth nerves, of severe degree. The face showed a weakness of the left side compared to the right. There was a left paresis of the upper motor neurone type.

He was very strong but the left limbs were less powerful and less well co-ordinate than the right. There was a few degrees limitation of supination of the left forearm and of the extension at the left elbow passively. Otherwise passive movements were full. In the left leg there was limitation of passive dorsiflexion of the ankle. The extent of voluntary movements could not be tested, but appeared to be little limited.

When made to move the left hand the finger movements were grossly inco-ordinate and there was marked slow separation and extension athetosis involving all the fingers and the wrist. He could grasp objects but not apparently release them.

The gait was grossly impaired with the left foot in a position of marked equinus and the hip tended to adduct so that the left foot was placed almost exactly in front and across the right. As he walked or when he used the right arm the left took up a position of adduction at the shoulder, flexion at the elbow, wrist and fingers and pronation of the forearm. There was a moderate increase of tone of the left limbs which were spastic. The biceps, triceps, supinator, knee and ankle jerks were much increased on the left compared to the right. There was no clonus. The Hoffmann signs were absent. The left plantar response was extensor, the right flexor.

Sensory findings were untestable except that pin prick was appreciated on the left limbs.

discarded within seconds in favour of some new feature of the environment which had captured the child's fleeting attention. Characteristically only running water would hold interest for more than a few seconds. The tendency to suck and chew was very marked in some cases and provided a major problem, since clothes, sheets and blankets, and even carpets were often destroyed.

Most of the parents complained of the children's failure to make any form of personal relationships and also that their children could be quite uncontrollably ferocious when frustrated, biting and scratching. Occasionally such ferocity might be apparent without any apparently justifiable precipitating factor. Punishment was ineffective in modifying the behaviour; phenobarbitone invariably exacerbated it, but a few patients did improve on regular amphetamine or primidone. In many cases spontaneous improvement occurs between the ages of 7 and 10 years.

Similar behaviour has been reported in epileptic children by Peterman (1953) and Ounsted (1955). A series of 25 patients with this form of behaviour, the majority of them suffering from cerebral palsy and half of them hemiplegic, was presented by Ingram (1956). Case 144 is a good example of overactivity in a case of acquired hemiplegia.

Other abnormalities.

A number of other abnormalities of clinical interest though of little importance from the functional point of view were encountered. A number of patients showed rather poorly developed hands and feet. These were small and the fingers tended /

tended to be poorly differentiated, being short and of rather similar length. The digits were frequently hyperextensible and syndactyly, especially of the toes, was common.

Two patients were born with bilateral talipes equino varus deformity and later acquired hemiplegic paralysis.

The relatively high prevalence of other abnormalities and the poor development of the hands and feet in patients with cerebral palsy was noted by a number of the earlier French and German authors. They suggested that they indicated the possibility of generally faulty development, which might be associated with increased susceptibility to neurological disease. (Féré, 1896; Freud, 1897).

One patient had a congenital dislocation of the hip on the hemiplegic side which defied all orthopoedic attempts to reduce it.

CHAPTER 5.a.

The Literature on Bilateral Hemiplegia.

The distinction between bilateral hemiplegia and other forms of cerebral palsy has been made gradually. Von Heine distinguished between congenital unilateral paralysis, or hemiplegia, and symmetrical (bilateral) paralysis. In symmetrical paralysis the lower limbs tended to be more affected than the upper whilst in unilateral palsy the upper limb was more severely paralysed than the lower. He noted occasional cases of symmetrical paralysis, however, in which the upper limbs were more affected than the lower (von Heine, 1860).

Little (1862) did not distinguish between cases of generalised rigidity and patients with bilateral hemiplegia, but did contrast conditions in which rigidity was the major cause of motor dysfunction with those in which paresis was the most striking abnormality.

Following the work of McNutt (1885) it seemed that it was unnecessary to make fundamental distinctions between the clinical syndromes of unilateral hemiplegia and generalised rigidity (termed bilateral hemiplegia by contemporary American clinicians). Both hemiplegia and "bilateral hemiplegia" were due to subdural haemorrhages sustained at the time of birth. The haemorrhages were unilateral in hemiplegia and bilateral in patients suffering from generalised rigidity or "bilateral hemiplegia" (Gowers, 1888; Lovett, 1888; Osler, 1889; Sachs and Peterson, 1890). Many of these authors considered that paraplegic rigidity was a different /

different clinical syndrome from generalised rigidity, because it was due to abnormalities in the spinal cord.

The first attempt to distinguish between patients in whom rigidity was marked, especially in the lower limbs, and children with paresis of movement more evident in the upper limbs than the lower was made by Feer (1890). He described patients suffering from hemiplegia who showed milder indications of paresis worse in the upper than the lower limb on the contralateral side.

More detailed criteria for distinguishing clinically between diplegia and bilateral hemiplegia were suggested by Dejerine (1892). In bilateral hemiplegia the upper limbs were more affected than the lower and there was almost invariably involvement of the cranial nerves. In diplegia ("generalised rigidity") the lower limbs were always more severely affected than the upper, and bulbar involvement was much less marked.

Freud (1893; 1897) also stressed the frequency with which cranial nerves were involved - "pseudo-bulbar palsy". He commented on the often marked weakness and lack of control in the trunk muscles and emphasised that these features made bilateral hemiplegia more than a summation of two unilateral hemiplegias. He noted that the upper limbs were more involved than the lower limbs by paresis, vasomotor changes, dwarfing and contracture than in generalised rigidity. Most of the patients were severely mentally defective. Head size tended to be small and epilepsy was very frequent. On the other hand combinations of diplegia and hemiplegia were occasionally /

occasionally encountered. Since they could be regarded as showing transitional forms between bilateral hemiplegia and generalised rigidity, it was probably wrong to think of bilateral hemiplegia and generalised rigidity as being completely different entities. He appears to have regarded bilateral hemiplegia as the most severe form of cerebral palsy in a sense comprising one end of a spectrum at the other end of which was mild paraplegia. This was also the view of Haushalter (1895). Neither of these authors gave much information about possible differences in the pathology of bilateral hemiplegia and other forms of cerebral palsy.

Since Freud's time the majority of neurologists have accepted his category of bilateral hemiplegia as comprising a usefully defined group of congenital bilateral palsies which the lower limbs are more severely affected than the upper (Perlstein, 1953). On the other hand others have made no distinction between bilateral hemiplegia and spastic tetraplegia ("diplegia") (Phelps, 1940; 1941; Wyllie, 1954; Evans, 1948). Very little has been written specifically about the condition though it seems to be accepted that it is the most severe form of cerebral palsy, often associated with severe mental defect, epilepsy and a poor life expectancy. The fullest recent account of the condition is by Ford. He emphasises that the pathological lesions are not uniform. Microgyria, pachygyria and various defects of development may be found, though the commonest underlying abnormality is true bilateral porencephaly. He admits that double hemiplegia may not always be easily distinguished from diplegia. In severely /

severely affected patients the symptoms are usually evident from birth but in milder cases may not be apparent for some months. "As a rule the patient is quite helpless and severely demented." Pseudo-bulbar palsy is nearly always present. Convulsive seizures are very common and mental defect is generally severe (Ford, 1926; 1952).

It is difficult to obtain any real idea of the frequency of congenital bilateral (double) hemiplegia or of the common aetiological factors or pathological changes from the literature. More detailed knowledge has to be derived from scattered case reports in which interest has usually been focussed more on the pathological than the clinical aspects of bilateral hemiplegia. It seems apparent that some of the patients described by Cohn and Neumann (1946) in their paper on porencephaly come into this category. The third case found to suffer from hydranencephaly described by Hamby et al.(1950) would also appear to have been a case of bilateral hemiplegia. The fuller clinical descriptions of Yakovlev and Wadsworth (1941) suggest that "schizencephaly" may underly the clinical picture in at least some cases.

A number of cases of bilateral hemiplegia with autopsy findings have been reported by Benda (1952) and Benda and Hoessly (1956). They emphasised the difficulties in determining the aetiology of many of the gross cerebral abnormalities which have been found in the condition. Though many of these findings must be regarded as being due to developmental aberrations originating at an early stage of gestation, others of them are more probably the result of birth injury. They describe /

describe in considerable detail a number of cases of cystic degeneration of the brain attributable indirectly to the effects of trauma which might have been considered to be developmental defects by other workers. The effect of pressure on the underlying growing brain when the meninges are unyielding following subdural haemorrhage is one example of the way in which these abnormalities may be caused.

In two papers by Yannet et al, a number of patients are described who were almost certainly suffering from bilateral hemiplegia but it is difficult to be sure how many there were. He describes the characteristic appearances of spasticity and contractures in flexion of all four limbs which had previously been hypotonic and paretic in a number of cases who were severely mentally defective, microcephalic and epileptic (Yannet, 1949; Yannet and Horton, 1952). Unfortunately his clinical groups are too varied, containing patients suffering from Tay-Sachs disease and tuberous sclerosis, for any reliable conclusions about bilateral hemiplegia to be drawn from his studies.

It is apparent that there is a great need for the collection of correlated clinical, aetiological and pathological findings in a large series of patients suffering from bilateral hemiplegia.

Aetiological Studies of Bilateral Hemiplegia.

Family history of patients with bilateral hemiplegia.

Rather a high proportion of the relatives of the 8% patients with bilateral hemiplegia appeared to be abnormal. The only abnormal father was in Case 92 who suffered from severe anxiety neurosis which was becoming worse the longer the grossly defective and impossibly overactive and destructive patient remained in the household. The only abnormal mother was in Case 95. She was said to have recently healed tuberculosis, but in no case was there a history of psychiatric mental or neurological disorder in the mother. (Table 68).

In Case 137 the older sibling had congenital bilateral dislocation of the hips. In Case 1 the father's sister died from convulsions at the age of 23. Another sister was a mental defective in the same institution as his daughter, and his uncle was said to be paralysed and defective. His nephew had a right hemiplegia, epilepsy and mental defect. In Case 92 the mother had a mentally defective cousin. In Case 137 one maternal aunt was said to be mentally defective and insane.

A full family history could not be obtained in Case 9, in which the child was illegitimate, or in Case 185 in which the parents were dead.

History of other pregnancies to mothers of patients with bilateral hemiplegia.

There was a total of 14 pregnancies to the mothers of children with bilateral hemiplegia, giving a fertility rate of 1.8 approximately, by far the lowest of any of the types of cerebral palsy. But the figure may be falsely low, for amongst the 8 patients one was illegitimate, and in two others the /

TABLE 68.

Family History in Bilateral Hemiplegia

Case No.	Social Class	Age of Mother	Number of patient's pregnancy	miscarriage	Other abnormal parturition	Other normal	Still-birth	Neonatal deaths	natal deaths	Abnormal siblings	Normal siblings
1	III	41	4 of 4	0	0	3	0	0	0	0	3
9	III	18	1 of 1	0	0	0	0	0	0	0	0
35	I	25	2 of 2	0	0	1	0	0	0	0	1
92	III	25	1 of 1	0	0	0	0	0	0	0	0
95	III	30	1 of 1	0	0	0	0	0	0	0	0
105	IV	33	2 of 2	0	0	1	0	0	0	0	1
137	III	24	2 of 2	0	0	1	0	0	0	1	0
185	III	21	1 of 1	0	0	0	0	0	0	0	0

the mothers were separated and unlikely to have further children.

Of the 6 pregnancies other than those resulting in the birth of the patients, none was abnormal, and only in Case 137 was the sibling resulting from the pregnancy abnormal, having dislocated hips.

It is impossible to work out any significant figures for the spacing of pregnancies when there are so few pregnancies with which to deal.

The birth histories of patients with bilateral hemiplegia.

The birth histories of the patients with bilateral hemiplegia are summarised in Table 69 . All the patients were classified as suffering from congenital bilateral hemiplegia except Case 105 in which there was a clear-cut history of normal development until the child sustained meningitis at the age of 7 months. In his case the birth was apparently quite normal.

Of the remaining 7 cases there was a history of normal pregnancy, labour and delivery in two (Cases 92 and 185). In five parturition was abnormal in some respect.

In Case 1 there was a prolonged second stage of labour of "more than a day and a half" and the child's shoulders became impacted after the head had been delivered. No doctor could be found for between 3 and 4 hours, and when one was obtained he had the greatest difficulty in extracting the child, who was born in a state of apnoea, shock and was intensely cyanosed. Regular spontaneous respiration was achieved only after about one hour of artificial respiration.

As in Case 1, pregnancy was apparently normal in Case 137, though /

TABLE 69.

Birth Histories of patients with Bilateral Hemiplegia

Case No.	Maternal age	Number of Pregnancy	Maternal health	Pregnancy	Labour	Delivery	Birth weight	Neonatal state
1	41	4	Obese but otherwise well.	Healthy	Prolonged 2nd. stage. Shoulder impaction.	Difficult extraction	7 lbs.	Prolonged apnoea. Very cyanosed. Fits for 1st. 2 days of life. Could not feed.
9	18	1	Well	Mod. well Pre-eclamp. Postmature 42 weeks.	6½ hours	Spontaneous vertex	7/7	Cried at once but very lethargic. Would not feed.
35	25	2	Healthy	Normal	Precipitate less than one hour.	Spontaneous vertex	6/13½	Cried at once. Feeding difficulty. Head retraction. Retarded milestones.
92	25	1	Healthy	Normal	14 hours	Spontaneous vertex	7/11½	Extremely lethargic. Noted to look abnormal.
95	30	1	Healthy	Antepartum haem. 5 days before deliv. at estimated 43 weeks.	60½ hours	Spontaneous vertex	7/13	Lethargic. Poor feeder. Convulsions from 3 weeks.
137	24	2	Healthy	Less life than last preg. otherwise well	5 hours foetal distress.	Spontaneous vertex	8/12	White asphyxia. Art. resp. & convulsions at 24 hours.
185	21	1	Healthy	Normal	Unknown	Unknown	7/4	Unknown
105	33	2	Healthy	Normal	"Few hours"	Spontaneous vertex	8/4	Normal

though the mother noted there was less "life" than in her earlier pregnancy. Foetal distress was noted during the apparently normal labour, and though spontaneous vertex delivery occurred, the child was delivered in a state of white asphyxia, almost pulseless. The cord was found to be only nine inches long.

In two cases the pregnancy was complicated. There was moderately severe pre-eclampsia in Case 9, but the child was successfully delivered spontaneously by the vertex after an apparently normal labour and cried at once and seemed normal initially. In Case 95 there was an antepartum haemorrhage five days before delivery. Labour occurred at an estimated 43 weeks' gestation and was exceedingly prolonged lasting with episodes of inertia for a total of 60½ hours. Delivery was eventually spontaneous by the vertex and the child cried immediately after delivery but was thereafter extremely lethargic and refused to feed.

The neonatal state and subsequent course of the patients with bilateral hemiplegia.

In Table 70. is summarised the details available about the immediate neonatal state and early course of the patients with bilateral hemiplegia. It will be seen that only two patients were delivered in a state of apnoea, the other five of the congenital cases (Case 105 being excepted from consideration) crying or breathing immediately after birth. Both these patients had convulsions within a few hours of birth. In Case 1 the convulsions were generalised and persisted for two days and were succeeded by a lethargic state in which the child refused /

TABLE 70.

Neonatal state and postnatal development in bilateral hemiplegia

Case No.	Parturition	Immediate neonatal condition.	Neonatal progress	Progress
1	Prolonged 2nd. stage Shoulder impaction. Difficult extraction.	Prolonged apnoea. Very intense cyanosis.	Fits for 1st. 2 days. Refused to feed and lethargic thereafter.	"Spasms" from 3 mths. to 1 yr. Very retarded milestones. Gradual stiffening of limbs. Epilepsy.
9	Moderately severe pre-eclampsia. Post-maturity 42 weeks.	Cried at once.	Extremely lethargic. Refused to feed. Difficulty swallowing.	Gradual stiffening of limbs at 6 months. Doubtful vision. Very retarded milestones.
35	Precipitate delivery at term.	Cried at once.	Lethargy, vomiting and feeding difficulty and head retraction in 1st. week.	Convulsions at 2 months. Retardation of milestones. Gradual stiffening of limbs at 6 months.
92	Normal.	Cried at once.	Lethargy. Looked odd, but fed well.	Convulsions aged 2 months. Retarded milestones.
95	Antepartum haemorrhage 5 days before delivery at estimated 43 weeks.	Cried at once.	Very lethargic. Poor feeding.	Convulsions at 3 weeks. Choking at 6 weeks. Gradual stiffening of limbs 3 mths. onwards.
137	Foetal distress during labour.	White asphyxia. Artif. respir.	Convulsions at 24 hrs. for 6 weeks.	Recurrence of convulsions at 3 months. Retarded milestones. Gradual stiffening of limbs from this time.
185	Probably normal.	Probably normal.	Poor feeding. Sleepy baby.	Lethargic. Retarded milestones. Fits aged 18 months. Gradual stiffening of limbs thereafter.
105	Normal	Normal	Normal	Had pneumococcal meningitis aged 7 months and seemed blind and unable to move thereafter.

refused to feed. In Case 137 they were generalised and persisted for 6 weeks and occurred many times a day. Between whiles the child slept and seemed uninterested.

In all cases there was a history of retarded milestones and increasing stiffness of the limbs. In Case 1 "spasms" (which in fact were generalised convulsions) recurred at the age of 3 months. Similar generalised convulsions of brief duration were noted at 2 months in Cases 35 and 92, at three weeks in Case 95, at 18 months in Case 185, and recurred at 3 months in Case 137. The only congenital case in which there was no history of convulsions was Case 9 in which insidious increase in the stiffness of the limbs was noted from the age of 6 months. Usually the convulsions were initially of the type called "myoclonic jerks" involving a sudden jerking flexion or extension movement of the limbs, neck and trunk so that the body is thrown into the foetal or an opisthotonic position for a few seconds with loss of consciousness. These "lightning major convulsions" (Buchanan) were present up to forty times a day for a period of months in Cases 35, 92, 95 and 137, and gradually diminished in the second year of life, being replaced by much less frequent grand mal convulsions in Cases 92 and 137. In Case 1 there was a gradual transition of the attacks into predominantly left-sided generalised convulsions at the age of about two years. In Case 95 there was a predominance of right-sided movement in the attacks from the start. In Case 35 there was a predominance of left-sided movement, especially marked in the tongue and face, and at times from the age of about 6 months attacks of focal /

focal twitching in the left face and tongue would be seen, without any jerking of the trunk.

Consideration of the birth histories of patients with bilateral hemiplegia.

In two cases of congenital hemiplegia the birth was apparently normal and the children seemed normal immediately after birth (Cases 92 and 185). Yet both babies were noted to be extremely lethargic, fed poorly and "looked odd". In Case 185 no relevant family history of mental, psychiatric or neurological disorder was obtained. In Case 92 the mother had a cousin who was mentally defective. Whether the family history is significant or not it does seem likely that both these patients were abnormal before they were born and it seems unlikely that manifest abnormalities of pregnancy or delivery were responsible. It seems probable that developmental defects may have been the cause of the patients' gross neurological abnormalities, though to what these abnormalities can be attributed, genetic causes, or accidents in early pregnancy, remains unknown. It is interesting that in Case 92 there was an associated congenital heart lesion which if due to the same aetiology as the cerebral defect would date the responsible insult to very early in pregnancy.

In the other five congenital cases (Cases 1, 9, 35, 95, 137) there were abnormalities of parturition confined entirely to late pregnancy, labour and delivery. To what extent these can be considered to have resulted in cerebral damage at the time of birth in the absence of neonatal apnoea seems problematical, especially when, as in Cases 35 and 137, other congenital abnormalities, of the hair-line in the former and the spine in the /

Case 35.

Girl, aged 4 years, with a very small head and a low hair line, suffering from bilateral hemiplegia with a history suggesting that birth injury may have occurred.

She was the second child of healthy parents. An older sister is normal. Pregnancy was uncomplicated. She was delivered at term in hospital after a labour of one hour, of which only 5 minutes were occupied by the second stage. She breathed at once but seemed lethargic after delivery. The birth weight was 6 pounds 13½ ounces. She was extremely difficult to feed, because of difficulty in fixing her to the breast and difficulty in keeping her awake during feeds once she was fixed. She vomited persistently shortly after feeds during the first week of life. Thereafter vomiting diminished in frequency but continued to occur in bouts of a few days every week. She did not smile and was unable to hold her head up by the age of two months.

At the age of two months she began to have generalised convulsions with loss of consciousness lasting for up to half an hour at a time and occurring several times a day. Between convulsions the head was noted to be retracted, and she refused to feed and seemed even more lethargic. She was admitted to hospital where the fontanelle was noted to be bulging and the presence of bilateral papilloedema was recorded. She was slightly febrile, the temperature varying between 99 and 100.5°F. Convulsions continued to occur. Lumbar puncture revealed straw coloured fluid containing 978 red blood cells and 65 white blood cells per cu. mm., of which 40 were polymorphs and 25 lymphocytes. The head circumference increased during the next 12 days, and she was transferred to a neurosurgical unit where the previous findings were confirmed and a head circumference of 15 inches was recorded.

An air encephalogram revealed the presence of a large spherical cyst high upon the right side of the brain just posterior to the coronal suture measuring 3 by 4½ by 4½ cm. This was tapped and produced thick yellow fluid which was considered to be the result of brain softening of some standing. The cyst connected with the subarachnoid space but not with the ventricle. The ventricular fluid, however, was also abnormal, containing 190 white cells per cu. mm. and 200 mgm. of protein.

The child continued to be apathetic and lethargic, but at the age of 16 months was noted to be attempting to sit with support and to be able to smile and laugh. She was still being fed from the bottle owing to her inability to swallow semi-solids. She spent hours at a time whining and crying and slept for no more than 1½ to 2 hours at a time. Continuous /

Continuous "smacking movements of the lips" were thought to be epileptic. These were not influenced by phenobarbitone medication, but whilst taking phenobarbitone no further major convulsions occurred.

She never learnt to sit independently, appeared to understand nothing of what was said to her and had gradually become very stiff and flexed in all four limbs. She never made any effort to reach with the hands, but laughed when spoken to and followed moving objects with her eyes. Intermittent smacking movements of the lips continued to occur.

It had never been possible to feed her with semi-solids on account of her inability to swallow. She had had an average of 2 - 3 severe respiratory infections, diagnosed by her doctor as "hypostatic pneumonia", each year. These had been successfully treated by sulphonamides, penicillin or broad spectrum antibiotics.

On examination. She was a small whining, helpless, miserable, defective child. At the age of four years her length was $36\frac{7}{8}$", her head circumference $18\frac{3}{4}$". There was a very low hair line anteriorly and posteriorly and the hair extended posteriorly from the occiput to the shoulders bilaterally.

She followed moving objects inconstantly but showed no appreciation of what was said to her though she appeared to hear, and reacted by jerking to sudden unexpected noises. She made no spontaneous voluntary movements with the limbs which were contracted in flexion. Vision was impossible to test fully. Fundi were normal. Pupils reacted normally. The face was expressionless and showed bilateral paresis, more marked on the right than the left. There was marked difficulty in swallowing, and tongue and soft palate moved sluggishly on stimulation.

There was a marked increase of tone of spastic type in all four limbs and these were held in positions of generalised flexion, the fingers being flexed over the adducted thumbs and the feet plantar flexed. The biceps, triceps, supinator, knee and ankle jerks were all grossly increased and the plantar responses were extensor. She reacted sluggishly to pin-prick in all regions of the body but detailed sensory testing was impossible.

The hands and feet were noticeably cold, and slightly blue bilaterally. Bilateral dislocations of the hips were suspected to be present but could not be confirmed by radiological examination in the circumstances of the survey.

the latter, were present. On the other hand it has to be recognised that the histories in both these patients are highly suggestive of birth injury. Case 35, in particular, was very fully investigated.

It seems probable that the material obtained on intracranial aspiration was cerebral tissue in a state of liquid necrosis and if this is so, it indicates the probability of fairly recent cerebral infarction, compatible rather with birth injury than with a developmental defect arising in very early pregnancy.

Similarly in Case 1. there is what amounts to a classical history of birth injury, with later apnoea, convulsions ensuing within two days and lethargic lazy behaviour thereafter. But the family history is suggestive of at least some predisposition to cerebral abnormalities on the father's side. It may be that a combination of hereditary predisposition and birth injury is necessary in these patients to produce congenital hemiplegia, but it is difficult to accept that all cases must be due to a combination of these factors. It seems much more likely that, as in other developmental malformations, parturition is more commonly disordered in cases with foetal cerebral maldevelopments than in those in which the foetus is normal (Murphy,1948).

Acquired bilateral hemiplegia.

In Case 105. there is no doubt that the child's development was apparently normal until he sustained an attack of pneumococcal meningitis at the age of 7 months which was not treated until about 10 days after marked irritability, neck stiffness, vomiting, refusing to feed, and fever first appeared. Very

gross cerebral destruction must be assumed to have taken place, presumably on a vascular basis as suggested by Cairns and Russell (1946). Children who survive with such gross neurological deficit as a result of meningitis are fortunately few, judging from the experience of this series.

The clinical findings in patients with bilateral hemiplegia.

Eight patients, one male and 7 females, with cerebral palsy were classified as suffering from bilateral hemiplegia. In all the impairment of motor function was paretic in type and more severe in the upper limbs than the lower. In all contractures were evident and all but one, who was considered to show moderately severe impairment, showed severe impairment of voluntary movement. All but one of the patients were bed-ridden.

All the patients were considered ineducable, either by the school authorities or the examiner. In none of the patients tested was the intelligence quotient more than 50. All the patients showed cranial nerve involvement and swallowing was difficult or impossible in six. Only one child showed any comprehensible speech. All but two of the patients were microcephalic, having a head circumference of less than 19" after the age of 4. The patients are similar to those described by Freud (1897) and were differentiated by him from those suffering from diplegia.

Motor impairment.

In all the patients with double hemiplegia the voluntary control of the limbs was deficient. It was difficult to gauge the severity of the deficiency because of the severe intellectual impairment, disused contractures and lack of co-operation shown by all the patients.

Only one of the patients was not classified as suffering from severe impairment of voluntary control of the limbs. She was a grossly overactive girl of 6, who was considered to be /

be moderately severely affected. She was capable of walking unsupported, though her gait was very spastic and unsteady. She was able to grasp with both upper limbs but did so in an infantile fashion, using all the fingers, not the thumb and forefinger, of the hands. Her handling was grossly clumsy. Release of the grasp was very poorly co-ordinated and objects were thrown from the hands rather than released from the grasp. Both hands were equally affected and marked athetosis of the fingers was evident. Full extension of both wrists and elbows and supination of the forearms was limited by contracture. The feet were incapable of passive dorsiflexion to the right angle owing to contracture and the hips showed some degree of flexion deformity. Associated movements of the upper limbs were marked and a flexion type when she walked. There was generalised spastic increase of tone more marked in the arms than the legs and rather more severe on the right side than the left, with marked bilateral exaggeration of the tendon jerks.

Of the 7 patients classified as suffering from bilateral hemiplegia of severe degree, all were bedridden and showed severe generalised flexion contractures of the limbs. None was able to feed himself. All were incontinent of urine and faeces. The degree of voluntary movement present in the limbs varied but was poor in all the patients. Only one seemed to take the least interest in attempting to handle objects. He was able to grasp them but not to release them from the grasp. Some of the other patients were able to grasp objects placed in their hands, but in all the grasp was achieved /

achieved by using all the fingers and not the thumb and forefinger. None was able to stand or to sit without support. Only one was able to sit even with support. Only three were able to hold up the head.

In all there was marked increase of tone in the limbs, more severe in the arms than the legs and predominantly spastic in type, though some rigidity, the result of contracture, was also evident. The tendon jerks were increased markedly in all. The degree of exaggeration of the tendon jerks was usually more marked on one side than the other. The side showing the greater exaggeration was not necessarily the more severely affected side, for in some contracture had limited the response of the muscle to the stretch stimulation.

In all the plantar responses were extensor, though whether the plantar response was of the infantile withdrawal type of a true Babinski response was sometimes difficult to determine.

Athetosis of the fingers, similar to that found in cases of unilateral hemiplegia was evident on attempted movement of the limbs in four of the cases of bilateral hemiplegia.

Dwarfing of the limbs.

The limbs, and especially the upper limbs, appeared to be small and generally poorly developed with reduction of girth, as well as of length, in all the patients, but because of the severe contractures in most, it was difficult to obtain accurate measurements. In only two cases was marked asymmetry of the limbs apparent. One patient showed apparent shortening of $1\frac{1}{2}$" in the upper limb and $\frac{3}{4}$" in the lower limb on one side compared /

compared to the opposite. Another showed 1" apparent shortening in the one upper limb and ½" shortening in the lower compared to those on the opposite side. It was interesting to observe in both these cases that the degree of contracture was approximately equally severe bilaterally.

Vasomotor changes in the limbs.

As in cases of unilateral hemiplegia vasomotor changes were evident in the limbs of those showing bilateral hemiplegia. In all cases the extremities were cold and the hands more severely affected than the feet. In three cases persistent cyanosis of the hands and feet was evident. In one of these cases there were marked chilblains of the hands and feet and a varicose ulcer was present over one tibia. This was similar in type to varicose ulcers of the adult.

Retardation of growth.

Unfortunately the number of cases is too small for the degree of growth impairment to be considered in a statistical manner, and since the time of onset of the bilateral hemiplegia was uncertain in some patients this would have been a matter of great difficulty, had it been possible.

All the patients were smaller than would have been expected if they had been normal children, however, and the apparent dwarfing appeared to be more marked in older than younger patients.

The head circumference.

The front occipital circumference was measured in all cases with bilateral hemiplegia and was found to be reduced, according /

according to the usually quoted figures, in all but two. All patients except these two had head circumferences below 19". In 5 the head circumference was 18½" or less, and in 2 it was less than 18". None of the patients was less than 3½ years at the time of measurement.

The shape of the heads varied greatly. Most cases showed a severe degree of occipital flattening.

One patient showed a head circumference which was in the lower part of the normal range. The one patient (Case 105) who had had cerebral thrombophlebitis complicated by meningitis in infancy, showed arrested hydrocephalus and a head circumference of 21¾" at the age of eleven.

Intellectual impairment.

All the five patients over the age of 5 years who were suffering from bilateral hemiplegia had been classed as ineducable by the education authorities. Of the three under the age of 5 years, all were grossly defective.

Only one patient had any comprehensible speech, and he could only imitate a few words which he had often heard repeated by his relatives or on the wireless. None of the other patients had any comprehensible speech and none was able to express his wants even in sign language. How much the patients appreciated of their environment was impossible to estimate. It was clearly very little.

Owing to the absolute lack of volition of most cases, contracture of even slightly affected limbs was inevitable. The one patient who had independent walking showed grossly overactive and disordered behaviour (Case 92). She was never still for a /

Case 92.

Girl aged 6 years with bilateral hemiplegia, epilepsy, mental defect and grossly overactive behaviour.

She was the only child of a father with progressively severe anxiety state and a healthy, though harrassed, mother who had a mentally defective cousin. Pregnancy, labour and delivery were uncomplicated. The birth weight was 7 pounds 11½ ounces. She cried at once but was noted to be very drowsy and difficult to feed on account of her persistently falling asleep in hospital. She was noted to be excessively hairy and to look "odd" by the ward sister.

She continued to be very lethargic on discharge from hospital but was successfully bottle fed and thrived. At the age of 14 weeks, by which time she was beginning to smile though she could not hold up her head, she had a generalised convulsion and was unconscious for more than an hour before being admitted to hospital "almost moribund". She rapidly recovered on oxygen therapy and no precipitating cause for the convulsions was found. Congenital heart disease was diagnosed.

Her development continued to be slow after discharge from hospital. She did not hold up her head until the age of one year, sat with support at the age of 2, and stood with support on her toes at 3 years. She said single words at the age of 3 but never achieved sentences or phrases. There was continual drooling of saliva.

Once she became mobile she was noted to be overactive. She never stopped roaming about her parents' flat and would handle object after object in the room, exploring them by touch, putting them to the mouth and discarding them after she had explored them in this way. She did not respond to rebuke or chastisement. She had an apparently uncontrollable urge to chew everything within reach and by the age of four it was almost impossible to keep her in clothes because of her desire to bite and chew what she was wearing. Apart from realising that fire hurt, she seemed oblivious to injuries and was continually becoming badly bruised through walking into objects and falling off pieces of furniture.

Her parents had never noted any constructive attempt to play with toys or to make anything with bricks. She merely sucked the latter and threw them about before proceeding to do exactly the same to something else. They thought that her vision was defective but that she could see quite small objects "if she wanted to". She had never achieved bladder or bowel control. She had attacks of crying with subsequent generalised convulsions of brief duration at night four or five times a month.

On /

On examination. She was a grossly defective, overactive girl with flexion contractures of all four limbs and very limited attention span. Her height was 39½ inches; head circumference 19½ inches. There was a prognathous deformity of the face, hyperteleorism, and a low hair line which extended on to the shoulder, like a mane posteriorly.

She was never still, running about unsteadily on her toes from object to object, handling them, sucking them and throwing them away clumsily. Often she would pick up discarded objects and treat them as if she had never seen them before. She paid attention to things for no more than a few seconds. She often ran into pieces of furniture, whether because of defective vision or lack of attention it was difficult to determine. When her mother attempted to control her over-activity she attacked her, hitting blindly but strongly, and with great violence. She appeared to understand little of what was said to her, but had a few single words used appropriately.

It was impossible to test her vision fully. Optic discs were very pale bilaterally. Pupils reacted only slowly to light and accommodation. There was slight nystagmus bilaterally. The face showed bilateral paresis. She reacted to loud noises. There was some prehension on the left but not on the right. There was spastic tetraplegia more marked on the right than the left with flexion contractures in all four limbs and markedly increased biceps, triceps, supinator, knee and ankle jerks bilaterally. The plantar responses were extensor.

She ran and walked without support but on her toes and unsteadily, holding her upper limbs flexed across her chest as she proceeded. Sensory findings were largely untestable, but she responded to pin-prick in all regions by attacking the examiner.

The heart was enlarged and there was a loud systolic murmur and thrill maximal in the third interspace to the left of the sternum. It was thought that she had a ventricular septal defect.

a moment, dashed clumsily about, falling frequently and damaging herself and all accessible furniture in her course. Her overactive behaviour was similar in type to that described when unilateral hemiplegia was discussed.

Visual impairment.

Vision was defective to some extent in all the cases of bilateral hemiplegia. Only 2 patients, those with hydrocephalus and limited speech, and with independent walking and gross behaviour disturbance respectively, showed any ability to detect more than the difference between light and darkness. The over-active patient appeared to have hemianopia and to have grossly impaired visual perception. She showed bilateral coarse nystagmus. The hydrocephalic patient was able to recognise doors opening and shutting and to follow the movements of people about his room.

One patient showed severe unilateral cataract. Four of the patients showed well marked bilateral optic atrophy.

Cranial nerve involvement.

All the patients with bilateral hemiplegia showed evidence of cranial nerve involvement.

Strabismus.

Six patients showed strabismus. In three there was unilateral abducent paresis. In one bilateral abducent was present. Two patients showed bilateral third nerve paresis, one with associated unilateral abducent paresis.

Facial./

Facial involvement.

Facial paresis was difficult to examine on account of the inability of the children to co-operate. Four patients appeared to have more or less symmetrical bilateral facial paresis of upper motor neurone type. Three patients appeared to have bilateral facial paresis more marked on one side than the other. Only one patient appeared to have paresis confined to one side of the face. Thus all the 8 cases showed either unilateral or bilateral facial paresis.

Hearing.

This was quite impossible to test in most cases as it was impossible to distinguish lack of comprehension from deafness and appreciation of vibration from actual hearing.

Swallowing.

In all cases a history of drooling and difficulty in swallowing was elicited. Difficulty in swallowing was still evident at the time of examination in all but two patients. None of these was able to take normal solid food, and their diet was comprised of slops. Drooling was evident at the time of examination in 4 of the 6 patients with difficulty in swallowing.

Owing to impairment of swallowing, much aspiration of food occurred in most cases. Five of the 8 patients with bilateral hemiplegia had a history of hospital admissions for respiratory infections on one or more occasions. It is probable that aspiration of food material was the most frequent cause of these. The patience of the mothers in the feeding of /

of these hopelessly defective children was quite unbelievably good.

The tongue.

It was impossible to test tongue movements in the patients with bilateral hemiplegia.

Epilepsy.

Grand mal epilepsy was present in 5 patients (Cases 1, 95, 105, 137 and 185). In three they had occurred from the age of three months or less (Cases 35, 95, 137), and in the other two had begun before the age of 18 months. In none of them were there obvious localising features. The frequency of attacks varied from between two to four a day, with loss of consciousness for as long as half an hour in each attack, to once a year. In all patients the effect of anti-epileptic drugs on the convulsions was disappointing.

A further two patients suffered from febrile convulsions, generalised in type without apparent localising features (Cases 35 and 92). In them grand mal convulsions without fever had not been noted, though in Case 35 there was more or less continuous involuntary movement of the tongue and lips which was considered to be an epileptic manifestation.

In only one patient was a clear cut history of myoclonic attacks in infancy elicited (Case 1). From the age of three months sudden extensor spasms of the trunk and neck with extension of the hips and knees, dorsiflexion of the feet, adduction of the shoulders, extension of the elbows, pronation of the forearms and flexion of the wrist and fingers, had been observed. /

observed. These attacks ceased at the age of about one year, when the first grand mal attacks occurred. Petit mal was not noted.

The level of consciousness of the patients with bilateral hemiplegia was conspicuously variable from minute to minute and from day to day. This made any assessment of intellectual level extraordinarily difficult.

Associated abnormalities.

Three patients, all female and showing microcephaly, had a curious hair-line posteriorly. It extended low down on the neck and tended to spread slightly over the trapexius muscles bilaterally. The hair was black and bushy, fine and curly in all three cases. Two of these patients showed excessive hair over the extensor surfaces of the upper limbs.

One patient showed moderately severe talipes equino-varus deformities of both feet which had been present from birth. One patient showed multiple skeletal deformities. Talipes equino-varus was present in one foot and valgus deformity of the other. There was a marked lumbar scoliosis with three hemivertebrae and bilateral congenital dislocations of the hips. One patient had a congenital heart lesion, thought to be interventricular septal defect.

One patient in an expensive private institution showed evidence of healed rickets.

CHAPTER 6.

THE LITERATURE ON DIPLEGIC PARALYSIS.

Whereas the early medical interest in hemiplegic paresis was shown by pathologists, clinicians studied diplegic patients before much was known of the underlying pathological findings. Indeed progress in understanding the aetiology of diplegia was slowed by the poverty of well authenticated pathological reports until after the turn of the century.

The first accurate case report which certainly describes a patient with diplegia was by an orthopaedic surgeon (Delpech, 1831). A girl delivered after an uncomplicated pregnancy and labour had generalised convulsions from the eighth day of life, and when examined at the age of 6 years showed generalised rigidity of the limbs, with grossly impaired co-ordination of voluntary movement. There was microcephaly and severe mental defect. Delpech thought that she had suffered from a disease of the spinal cord and that the brain, which he believed developed from the cord, had failed to develop fully.

The first large series of children suffering from diplegia was presented by Little (1843, 1862). He described 63 cases of cerebral palsy in this latter paper, the majority of whom showed paresis of voluntary movement associated with rigidity in the limbs. The rigidity and paresis might be paraplegic in distribution or affect the trunk and both upper and lower limbs. The increase on tone in the limbs was more marked in the lower limbs than the upper in all cases, and walking was achieved very late in the majority. The rigidity was usually exacerbated by the patients being placed in the erect position with /

with their feet on the ground. "Scissoring" of the legs due to adductor spasm, a tendency to forced extension at the knees and plantar flexion of the feet then occurred. In other cases there was a marked tendency to flexion contracture of the limbs. Dwarfing of the lower part of the body and the lower limbs was frequently found. Little noted that the heads of the majority of patients were small and that strabismus was common. Speech was impaired more frequently and severely in patients with upper limb involvement than in those in whom the legs only were affected. Epilepsy occurred in a high proportion of patients. Though mental defect was frequent the defective appearance of patients at first sight was often misleading. Little was particularly concerned with the treatment of the secondary deformities found in diplegic patients, especially the correction of the talipes equino varus deformity of the feet which was so frequently found.

His papers are of particular importance because of their attempts to relate the causes of cerebral palsy to preceding abnormalities of parturition. The most important causes of damage were abnormal presentations of the child, difficult vertex deliveries often due to rigidity of the maternal soft parts, version, breech delivery and torsion of the cord. Premature birth was also found frequently. These abnormalities of parturition became know as "the factors of Little" (Freud, 1893). Their most frequent mode of action was to cause spontaneous respiration to be delayed. The consequent asphyxia produced great vascular congestion and small haemorrhages throughout the meninges, brain, brain stem and

spinal /

spinal cord. He emphasised that failure to breathe after delivery was associated with later rigidity in only a very small proportion of cases - "the majority wake up unharmed". He also presented cases in which no history of birth injury preceded the appearance of symptoms.

Though cerebral palsy was discussed only as a differential diagnosis of spinal paralysis in children by von Heine (1860), his clinical descriptions of generalised and paraplegic rigidity are the equal of Little's about whose papers he apparently knew nothing. Because of the frequent history of forceps delivery and the high proportion of patients showing upper limb involvement, mental defect, epilepsy and strabismus, he suggested that both paraplegic and generalised rigidity were more likely to be caused by cerebral than cord damage. This opinion was shared by Benedikt (1868) so far as generalised rigidity was concerned, but he thought that "paraplegic rigidity" was more readily explained as being due to spinal cord pathology.

This opinion was also held by Charcot (1884). He had recently described the findings in amyotrophic lateral sclerosis and stated, "It has never been possible to demonstrate a case of paraplegic rigidity in which a cerebral cause of the disorder could be shown." "Tabes spasmodique" became generally accepted by his scholars as being spinal in origin and was considered to be due to agenesis or atrophy of the long tracts. It was suggested by Brissaud (1894), for example, that if premature birth occurred at the seventh month, then the budding long tracts were affected early and generalised rigidity /

rigidity would result, whereas if they were affected later the arms might be innervated but the tracts would be affected before they had had time to reach the legs which would therefore be rigid. This point of view was supported by Marie and as late as 1903 by Dejerine. A similar opinion had been held by Erb (1877). But the majority of German, British and American workers believed that the condition was primarily cerebral in origin.

The similarity of the clinical features of generalised and paraplegic rigidity, the occurrence of mental defect and convulsions in both types, and the aetiological factors which were common to both were pointed out by Seeligmüller (1879). He also differentiated amyotrophic lateral sclerosis and multiple sclerosis of cord origin from the paraplegic and generalised rigidity in children which appeared to be of cerebral origin. That the failure of long tracts to develop might be the result of cerebral and medullary lesions was suggested by Naef (1885). In Britain the cerebral origin of generalised and paraplegic rigidity was emphasised on clinical grounds by Ross (1883) and Gowers (1888).

In the meantime a few autopsies of patients who had suffered from paraplegic and generalised rigidity had been reported in Europe. These showed a great variety of pathological appearances in the brain (Freud, 1893). Much greater attention, however, was paid to the well publicised work in America of Sarah McNutt (1885). She described the condition of generalised rigidity which she termed "bilateral hemiplegia" and attributed it to the presence of bilateral subdural effusions

the result of traumatic delivery. Her evidence for doing so was slight and her work was dismissed somewhat laboriously but thoroughly by Freud, and more abruptly by Collier (1899, 1924). They pointed out that the patients whose autopsies she reported had not suffered from generalised rigidity and that the patient with generalised rigidity had not been shown to have subdural haemorrhages. Nevertheless her opinion was widely accepted and did serve to emphasise the cerebral origin of the condition to American clinicians (Osler, 1888; Sachs and Peterson, 1889; Gowers, 1888).

Further advances in the understanding of the underlying processes in generalised and paraplegic rigidity were handicapped by the rather small number of autopsies reported, but with the increase in interest aroused by McNutt's articles more reports became available. Bilateral porencephaly was recorded by Otto (1885), atrophic lobar sclerosis was observed by Henoch (1892), Bourneville and Pilliet (1891) Friedman (1893). A case in which cerebral development appeared to have been interrupted at about the third month of gestation was described by Schultz (1886). Generalised shrinkage of the brain with thickened dura was reported by Peterson (1894). These and other pathological reports were collected by Freud (1893, 1897).

Freud analysed a high number of cases from the literature and from his own experience. He coined the term diplegia which included four types of neurological disorders occurring in early childhood which would be defined in clinical, and to some extent aetiological, terms, though intertypes between all disorders occurred. His categories were:- generalised rigidity /

rigidity, paraplegic rigidity, bilateral hemiplegia, and bilateral athetosis. It was necessary to describe each type separately, but the aetiological, pathological and clinical differences between generalised and paraplegic rigidity were in degree rather than in kind. He pointed out that the underlying pathology in generalised and paraplegic rigidity was very diverse in different cases and that it was almost impossible on the basis of the clinical history in a given case to guess at what particular pathology was present.

He attempted to assess the relative importance of the various aetiological factors which had been suggested on the basis of 270 cases taken from the literature. The cause was unknown in 34%, prenatal abnormalities were present in 7.7%, mixed prenatal and natal or natal and post-natal factors were present in 13%, extrauterine in 7.7%, and "Little's factors" were present in 50%. Other authors found "Little's factors" to be much less important. In 53 personal cases the cause was unknown in 42.7%, maternal factors were important in 31.4%, "Little's factors" in 9.2%, and extrauterine causes were operative in 16.6%, according to Ganghofner (1895, 1896). Freud (1897) suggested that the differences between series were probably due to the fact that cases tended to be published more often when some antecedent abnormality in the history could be suggested as a cause than when it could not. The danger of assuming that abnormalities of parturition were necessarily responsible rather than concomitant with cerebral abnormalities in the child was stressed by Freud. Apart from the fact that the pathological material at his disposal was rather inadequate and /

and his knowledge of the newborn academic, Freud's contribution was remarkably complete, and it is hardly surprising that rather few new ideas have been added since.

In the next ten years attention was concentrated more on cases of acquired diplegia and the types of diplegia which become worse in post-natal life (Sachs, 1891, 1892; Spiller, 1898).

More recent studies.

Since the turn of the century study of diplegia has been handicapped by differences in the terminology and classification of cerebral palsy. Whilst American authors were studying "bilateral hemiplegia" which included patients suffering from diplegia, authors on the continent were investigating children suffering from "diplegia" which comprised "paraplegic rigidity", "generalised rigidity","bilateral hemiplegia" and "bilateral athetosis". More recently "spastic cases" have been investigated. In some instances this term has been used to describe children with hemiplegia as well as those suffering from symmetrical paralysis of the limbs worse in the legs than the arms (Latham, Anderson and Eastman, 1954). In other series children suffering from progressive paralysis are included with those showing static congenital paresis (Collier, 1924; Penrose, 1938; Patten, 1924). It is necessary to scrutinise recent reports with some care before assuming that the findings reported in them are comparable with those in patients suffering from diplegia reported in this series.

STUDIES OF AETIOLOGY.

Pathological studies. /

Pathological studies.

Impressed by the reports of patients with symmetrical paralysis of the limbs dating from birth or shortly afterwards who deteriorated rather than improved as they matured, Collier considered that "diplegia" was progressive in a proportion of cases (Sachs, 1891, 1892; Spiller, 1898; Collier, 1900, 1924). He regarded diplegia as a collection of conditions due to primary neuronal degeneration. The common pathological finding in them was that they showed "killed, cut, stunted and retarded neurons". The neuronal degeneration might occur before or after birth and might become arrested or be progressive. Amongst his cases were some suffering from Tay-Sachs disease.

"I would put the following conditions forwards in explanation of the universal affection and of the various local affections of the brain which are found in diplegia. 1) The severity of the cause which may affect all the cerebral elements indiscriminately, if acting early giving rise anatomically to a small primitive type of brain destitute of neurons and causing clinically a condition of idiocy with diplegia and blindness. 2) The time of development at which the cause is active which may determine those systems which are affected and those which are spared in the local atrophies of the brain. 3) The physiological selective capacity of the cause to affect some systems and not others. 4) A cause which may be temporary in its action and which for this reason only picks out systems which are vulnerable at the moment. 5) A cause which may be continuous in its action and which results in a progressive diplegia."

He /

He regarded the disorder as being due to "hereditary and familial influences" in a proportion of patients and minimised the significance of birth injury. "Abnormalities of birth are not causal factors but common, though by no means constant, concomitant factors, indicative of something wrong with the foetus or the mysterious relationship between mother and offspring that makes for speedy delivery at term. My authority in the inconstancy of these so-called factors, the insuperable difficulty of referring one and the same pathological cause to prematurity, precipitate and prolonged labour and asphyxia, and the absence of any pathological findings to show how events might have been precipitated." Similar views were expressed by Kinnier Wilson (1954).

Patten (1924) thought that many of the pathological appearances could be explained on the basis of prenatal ischaemia. But he also noted the possibility that "developmental defect or arrest which concerns either the integrity of the cortical cells or the proper myelinisation of the corticospinal tracts or associated fibres" might cause the disorder. Unfortunately it is clear from the clinical descriptions of his 46 cases that some suffered from dyskinesia rather than diplegia and it is probable that others would have been classified as "ataxic diplegics".

A series of 50 autopsies on "diplegic" patients, the majority of whom had been severely mentally retarded, was reported by Stewart (1942). As in Patten's series, there were included cases which would not have been classified as being diplegic in the present survey. However, the majority of patients /

patients would probably have been included in this category. Normal pathological appearances were noted in 9 cases, atrophic lobar sclerosis in 6, microcephaly in 16, microhydrocephaly in 3, cortical atrophy without obvious sclerosis in 6, markedly abnormal convolutional pattern in 13, cerebellar atrophy in 5, developmental anomalies in one, optic atrophy in 2, small pyramids in 12, thickening of the pia-arachnoid in 13. He emphasised the diversity of the pathological appearances and was unable to suggest which aetiological factors might have been important even in many of the individual cases.

Many of the appearances which were described by Stewart have been produced artificially in experimental animals subjected to perinatal hypoxia (Windle and Becker, 1944). Partly as a result of these studies and of observations made in large numbers of patients dying at different periods after hypoxia, a clearer picture has emerged as to what pathological appearances are produced by perinatal hypoxia. Lobar sclerosis, for example, is regarded as being almost invariably due to cerebral hypoxia (or ischaemia) by Courville (1953). That cystic degeneration of the brain may often be the result of severe hypoxia or to disturbances in the cerebral circulation, rather than to developmental malformation, as previously believed, has been suggested by Benda and Hoessly (1956).

It seems clear from the increasing number of pathological studies of diplegic patients which have been reported that developmental malformations and perinatal brain injury both cause diplegia. The main argument centres on the size of their respective contributions and the proportion of developmental malformations /

malformations which are genetically determined (Minkowski, 1952; Courville, 1958).

Clinical Studies of Parturition.

A number of recent studies have demonstrated an apparent paradox; that the proportion of diplegic patients born prematurely is higher than the proportion of patients suffering from other forms of cerebral palsy, yet parturition is less frequently abnormal otherwise in diplegia than in the other types.

Evans (1948) found that amongst 38 patients suffering from "spastic para- or tetra-plegia" of congenital origin, there was a high prevalence of premature birth. Complications of pregnancy and labour were not significantly more frequent than in a control group, nor less frequent than in a group of "athetoid" patients. On the other hand, the proportion of "Athetoid" patients who suffered from asphyxia after birth or required resuscitation was higher than the proportion in the "spastic" or the "control" groups.

Asher and Schonell (1950) were impressed by the fact that whilst "athetoids" had a more frequent history of abnormal labour than patients with "symmetrical quadriplegia", the latter had a much higher incidence of premature birth. They suggested that the high proportion of males amongst patients with "spastic paralysis" was in favour of "a congenital defect in many cases, for most congenital defects have a predilection for one sex rather than the other". They found that the frequency of birth injury was no less amongst patients with "symmetrical para- or quadri-plegia" than amongst patients with /

with hemiplegia. They noted that this was not in accordance with the findings of other authors (Ford, 1926; McGovern and Yannet, 1947).

Sixteen of 75 patients suffering from quadriplegia were noted by Wood (1957) to have been born after apparently normal parturition. Some of these patients acquired their cerebral palsy post-natally, and some almost certainly would have been diagnosed as suffering from bilateral hemiplegia in the present survey. In a group of 26 patients diagnosed as suffering from paraplegia, of which 22 were probably cases of cerebral palsy, 15 had been delivered prematurely. Of the 86 patients in this group who would have been considered as suffering from congenital diplegia on the information provided, 26 (or approximately 30%) were born after uncomplicated parturition. Patients suffering from "dyskinesia" or "hemiplegia" had a history of abnormal parturition much more frequently.

Similar findings were reported by Salmonsen and Skatvedt (1955). These authors noted that 50% of children suffering from bilateral spastic paralysis (excluding mixed forms) were prematurely born compared to 17% hemiplegic and 19% "athetoid" patients. Whereas 48% of hemiplegic patients had a history of abnormal labour, only 34% of patients with "symmetrical spastic paralysis" had such a history.

The association of "diplegia" with premature birth has been noted by a number of recent authors. One third of 30 patients suffering from "spastic tri- or tetra-plegia" (probably including patients suffering from ataxic diplegia though not patients with bilateral hemiplegia) were noted to be /

be premature by weight in the series of Eastman and DeLeon (1955). Half of their 18 patients suffering from paraplegia were immature at birth. Childs and Evans (1954) observed that whilst the mean weight of patients suffering from "spastic tri- or tetra-plegia" was 5.97 pounds and their patients contained an excess of prematurely born children, the mean weight of paraplegic patients was still lower - 4.73 pounds - and the proportion of prematurely born babies was higher. There was a very high standard deviation in the weights of both groups compared to normal babies. There was a marked deficiency of "diplegic" patients weighing about 8 pounds, compared to the normal population. The distribution of diplegic patients by weight was remarkably similar to the distribution of "non-survivors" of the neonatal period.

The prevalence of diplegia amongst babies born in Edinburgh between 1948 and 1952 was found to be 4.6% amongst children with a birth weight of 4 pounds or less, but less than half this amongst those with birth weights between 4 pounds and 5½ pounds (Ingram and Kerr, 1954). These authors compared the aetiological factors common to diplegia and retrolental fibroplasia which accounted for their coexistence in 6 prematurely born patients. They found a very high prevalence of abnormal pregnancy compared to unaffected children or patients who suffered from only one of the conditions. "Hypoxic insults" occurred significantly more frequently in the perinatal histories of patients showing both retrolental fibroplasia and diplegia than in those showing only one of these conditions, and more frequently in children showing either retrolental fibroplasia /

fibroplasia or diplegia than in mature diplegic patients or in apparently healthy prematurely born babies. Nevertheless it is worth noting that of the 11 prematurely born diplegic patients, 3 had had apparently normal pregnancies, 4 normal deliveries and the condition of 2 children immediately after birth was described as good.

Churchill (1958) attempted to distinguish patients suffering from uncomplicated diplegia from those in which classical "Little's disease" was complicated by athetosis, ataxia or other aberrant neurological abnormalities. He found that patients suffering from "simple, spastic diplegia" were far more frequently the result of premature birth than were those with complications. He interpreted these findings as indicating that "simple spastic diplegia" had a specific association with low birth weight. It seems likely that by his rigid exclusion of patients showing other than "pyramidal signs" he avoided the tendency shown by most other authors to include cases of dyskinesia, bilateral hemiplegia and ataxic diplegia in his series. It is noteworthy that patients suffering from dyskinesia, ataxic diplegia and bilateral hemiplegia almost invariably show more marked upper limb involvement than do patients suffering from diplegia. It seems likely that other authors have diluted their cases of "spastic tri- or tetra-plegia" with patients suffering from these disorders. This would explain why they find a higher proportion of prematurely born patients suffering from paraplegia than they do of "spastic triplegia or tetraplegia". Not all the latter cases suffer from diplegia, the majority of the patients they term "paraplegia" do.

Multiple /

Multiple Birth. It does not seem possible to explain the high proportion of diplegic patients born prematurely on the basis of there being a higher prevalence of abnormal pregnancy than is found in other forms of cerebral palsy.

Some of the excess of prematurely born patients may be due to the relatively large number of diplegic twins. Evans (1948) found that 4 of 38 cases of "spastic paraplegia or tetraplegia" were twins, and Asher and Schonell (1950) found 8% of patients with "symmetrical quadriplegia" and 3% of patients with "asymmetrical spastic quadriplegia or paraplegia" to be twins. The proportion of twins was higher in diplegia than in hemiplegia or dyskinesia in both these studies and in those of Greenspan and Deaver (1953) and Shyh-Jong (1955). But it is clear that the major part of the excess of premature births cannot be attributed to the greater frequency of multiple birth in diplegia than in other forms of cerebral palsy.

Developmental Malformations. Other explanations of the high proportion of prematurely born diplegic patients include suggestions that the patients are delivered prematurely because they are congenitally abnormal, most often due to developmental malformations. The relatively high proportion of diplegic patients suffering from other congenital anomalies found by some authors has been held to support this idea. Diplegic patients suffering from Sturges-Weber's disease, aplasia of the corpus callosum, cleft palate, spina bifida, osteogenesis imperfecta and polydactyly, have been described by Andersen (1954) and Asher and Schonell (1950). Whether the prevalence of associated congenital malformations is significantly greater amongst diplegic patients than amongst the general /

general child population awaits statistical proof, though the figures of Eastman and DeLeon (1955) suggest that this is so.

The fact that diplegia appears to be familial more frequently than other forms of cerebral palsy is also held by some authors to indicate that a proportion of cases are due to developmental malformations which may be genetically determined in a proportion of cases (Penrose, 1938). Two siblings were found to suffer from symmetrical spastic paralysis amongst the 167 cases studied by Asher and Schonell (1950). "Spasticity" was noted in more than two offspring in three instances amongst 186 patients studied by Andersen (1955). Congenital paraplegia was noted by Woods (1957) in two sisters, and in patient, mother, grandfather, great-uncle and two uncles. Five of 7 severely mentally defective patients gave a history of siblings of blood relatives being similarly affected, and 2 of 65 patients suffering from "diplegia" (by which was meant more or less symmetrical paresis of the limbs involving more than the legs) were siblings (Yannet, 1949).

It is certain that in a number of patients reported as suffering from diplegia the disorder has been genetically determined. Families in which congenital spastic paraplegia was present in more than one member have been reported by Phillip (1949) and Dick and Stevenson (1953). In the majority of cases of familial paraplegia, however, the condition appears during childhood or in early adult life, as in the three siblings described by Sutherland (1957). It is difficult to guess on the basis of current information what proportion of cases of diplegia are genetically determined. There appears to /

to be no doubt that the familial prevalence of diplegia is higher than that of most other forms of cerebral palsy in published series. It seems possible that the rather high prevalence of siblings who have died or are abnormal though not suffering from cerebral palsy may also have some genetic significance (Penrose, 1938; Yannet, 1949; Illingworth, 1958). The majority of recent authors still consider that "anoxaemic brain necrosis occurring during the birth period" must be held responsible for diplegia in the majority of patients (Salmonsen and Skatvedt, 1955).

Acquired Diplegia.

Occasional cases of acquired diplegia clinically indistinguishable from the congenital variety have been recorded by a number of recent authors. Cases due to accidental hypoxia have been recorded by Courville (1953), after nitrous oxide anaesthesia by Ford (1952). It has been found by Radermacker (1956) after acute disseminated encephalomyelitis associated with a variety of infections. Benda (1956) has noted patients becoming paretic and spastic in all four limbs following trauma, in whom bilateral "cystic disease of the brain" was found post mortem. It is doubtful if either of the patients suffering from paraplegia after meningitis recorded by Woods (1957) were suffering from cerebral palsy, and at least four, and possibly more, of her 7 cases of "acquired spastic quadriplegia" would have been classified as suffering from bilateral hemiplegia in the present series. Nevertheless it seems clear that in a small proportion of diplegic patients the condition is due to post-natal disease.

Studies /

Studies on Clinical Aspects of Diplegia.

Neurological Findings. A number of authors have studied the neurological findings in infants and young children in the last forty years, but their importance in cerebral palsy has only gradually been appreciated. Symmetrical and asymmetrical neck reflexes, body righting, falling, stepping and startle responses of different types may be observed in normal children at various stages of their development (Magnus, 1926; Zador, 1938; Thomas and Dargassies, 1952; Koupernik, 1954). Their presence or absence in children suffering from cerebral palsy, especially dyskinesia or diplegia gives clear indications of levels of neuromuscular maturation and the extent of brain damage in many cases which cannot be gauged by other criteria. As well as being of diagnostic importance, these reflexes, the majority of which are medicated entirely by subcortical pathways, may also be used in therapy (Temple Fay, 1953; Bobath and Bobath, 1950, 1953, 1956). For example, flexion of the head may be utilised as a manoeuvre to reduce rigidity of the limbs in diplegia and the severity of involuntary movement in many patients suffering from dyskinesia. Close study of the neurological findings in children suffering from diplegia at different ages has resulted in an increased appreciation of the changes which may be observed in the manifestations of diplegia as patients mature, and as their subcortical reflexes become progressively inhibited by the activity of higher centres in the nervous system.

A detailed account of the neurological findings in maturing patients suffering from diplegia has been presented. It is possible (arbitrarily), to distinguish stages in the development /

development of the condition which may be closely related to the stages observed during maturation in normal children. Initially patients tend to show poverty of volitional movement, by hypotonic and show little response to changes in posture. Gradually this stage is succeeded by one in which postural behaviour is dominated by the symmetrical and asymmetrical neck reflexes. Sudden back arching movements, accompanied by extension of all four limbs and increased tone in the anti-gravity muscles, occur whenever the child's head is extended or he is placed in the erect position. These sudden extensor movements, which may be termed dystonic attacks, result in the patients transiently assuming positions of opisthotonos. The period during which this occurs frequently may be termed "the dystonic stage". It is succeeded by a period of weeks, months, or years, during which there is persistent increase of tone in the limbs, especially in the antigravity muscles, and a tendency for "rigidity" of the limbs to be marked. The limbs and trunk tend to be maintained in constant positions of extension, though this position becomes more extreme and rigidity (antigravity hypertonus) more marked when the child is in the erect position or there is pressure on the soles of the feet, or the head is extended. The period during which this constant rigidity in extension may be demonstrated was termed "the rigid phase" of diplegia. It in turn is gradually succeeded by a period during which flexed positions of the limbs begin to predominate over those of extension, and true spasticity with increased tone in the flexor muscle groups, stretch responses and increased biceps, triceps, supinator, knee and ankle /

ankle jerks may be demonstrated. Flexion contractures are likely to occur. This may be termed "the spastic phase" of diplegia. All these affected patients suffering from diplegia will continue to have "dystonic attacks", and functional use of the hands will always be rudimentary. Less affected patients may remain "rigid" for the rest of their lives, and though they will have better manual dexterity than those showing continued dystonia, prehension is unlikely to be achieved. Prehension is achieved in many patients in the spastic phase of diplegia and their level of neuromuscular function more nearly approaches the normal than that of children with dystonia or rigidity. Though all patients suffering from diplegia do not show these characteristic stages, their recognition has made early diagnosis of the condition easier, and prognosis can be more soundly based on the results of neurological examination than previously (Ingram 1955, 1959). A fuller account of the changes in neurological findings which occur in diplegic patients will be presented elsewhere in this work.

There is still some difference of opinion as to whether paraplegic, triplegic and tetraplegic patients should be considered in one category or separately. The majority of recent authors consider that patients suffering from "spastic para-, tri- or tetraplegia" show differences in the extent and severity of their neurological disorder rather than differences in kind (Evans, 1948; Asher and Schonell, 1950; Ingram, 1955). On the other hand a few still prefer to consider paraplegia and tetraplegia as different categories of cerebral palsy (Woods, 1957).

A number of recent specialised studies have added to the knowledge /

knowledge of other neurological abnormalities in diplegia. For example, the causes of strabismus have been analysed and classified according to whether there are local, muscular abnormalities, lower motor neurone paresis, or evidence of suprabulbar involvement (Guibor, 1954). The same author has described the frequency with which severe refractive errors, most commonly myopia, are encountered. The occurrence of retrolental fibroplasia in diplegic patients was noted by Ingram and Kerr (1954) and Eastman and De Leon (1955).

Strabismus was observed in 33% of patients suffering from "spastic quadriplegia" in the series described by Asher and Schonell (1950). Skatvedt (1958) noted refractive errors in 12 and strabismus in 25 of 121 patients with "bilateral spasticity". "Muscular imbalance of the eyes" was found by Woods (1957) in 9 of 26 paraplegic and 40 of 75 "quadriplegic" children. "Impaired sight" or blindness occurred in one paraplegic and 21 quadriplegic patients. She felt that the "impaired sight" was more often due to cerebral than local causes.

A number of rare, genetically determined congenital diplegic syndromes have been recognised. The association of oligophrenia, congenital icthyosiform erythrodermia, and maculoretinal degeneration was noted by Sjøgren (1956) and Sjøgren and Larsson (1957). Cases have been recorded complicating phenyl pyruvic oligophrenia and tuberous sclerosis (Woods, 1957; Andersen, 1954). As already described, a number of apparently uncomplicated congenital familial cases have been described (Dick and Stevenson, 1953).

Orthopaedic /

Orthopaedic surgeons have made a large number of studies of the deformities secondary to muscular spasm which may result in diplegia. The frequency with which fixed positions of talipes equino varus or valgus, flexion of the hips and knees, pronator contracture and flexion contracture of the elbow and wrist occur has been described. Secondary dislocation of the hip has been realised to occur much more frequently than used to be believed and is no longer confused with the congenital variety (Watson Jones, 1926; Matthews, Jones and Sperling, 1953; Tachdjian and Minear, 1956). Reviews of progress made in the recognition and treatment by surgery of secondary deformities in cerebral palsy have been presented by Cooper (1950), Carroll and Craig (1951), Baker (1955), Pollock (1955), Green and McDermott (1957), Phelps (1957) and Pollock and Sharrard (1958).

Epilepsy. The prevalence of epilepsy in diplegia has been variously estimated in different surveys, but the majority of recent reports suggest that seizures occur less commonly in diplegic patients than in those suffering from hemiplegia, and more commonly in those with dyskinesia. Asher and Schonell (1950) found that 23% of "spastic quadriplegics" had seizures compared to 3% of "spastic paraplegics". A higher proportion of patients with upper limb involvement than with only lower limb involvement were found by Perlstein, Gibbs and Gibbs (1955) to have convulsions. These authors found that 33% of paraplegic patients had seizures, which may be compared to the figure of 19% for paraplegics and 49% for cases of "spastic quadriplegia" given by Woods (1957).

Skatvedt /

Skatvedt (1958) studied the convulsions and electro-encephalographic findings in patients with cerebral palsy in some detail. She found that seizures occurred in 34.8% of 89 patients suffering from hemiplegia and in 19% of 121 cases of "bilateral spasticity". Epileptogenic foci in the absence of overt seizures were found on the encephalogram relatively more frequently in patients with hemiplegia than in patients suffering from "bilateral spasticity". A higher proportion of hemiplegic patients suffered from focal seizures than patients with bilateral spasticity, but the latter more frequently had atypical seizures, such as myoclonic jerks and akinetic seizures. The response of hemiplegic patients to anti-epileptic medication was less good than that of patients with bilateral spasticity.

Though Skatvedt felt that "EEG has proved a valuable supplement to the diagnostic armoury. In many cases it provided information with regard to the extent of lesions, which is not otherwise obtainable", the experience of other authors is that electroencephalography provides information of relatively little value in bilateral cerebral palsy. Perlstein (1955) observed that irrespective of the electro-encephalographic findings the seizures in cerebral palsy tended to be more resistant to anti-epileptic control than other forms of epilepsy.

Psychological Aspects.

A number of surveys of the intelligence of patients suffering from diplegia have been made in recent years. Unfortunately they cannot be compared directly to the results in /

in the present series or, in many cases, to each other, for very different classifications of cerebral palsy are used. The majority of surveys include patients suffering from bilateral hemiplegia and ataxic diplegia (who tend to be severely mentally defective) with diplegic patients in categories of "bilateral spasticity" or "spastic quadriplegia".

The majority of authors have found that spastic children (patients suffering from hemiplegia or bilateral spastic disorders) are less intelligent than those with dyskinesia (Phelps, 1941; Evans, 1948; Floyer, 1954). This is contrary to the findings of O'Brien (1945) and Asher and Schonell (1950).

The latter authors found the mean intelligence quotient of 286 "spastic" children to be 67.9 and of 45 "athetoid" children 67.6. There was a higher mean intelligence quotient in 87 patients suffering from "spastic paraplegia" (74.3) than in 84 children with "spastic quadriplegia" (50.2). When all patients suffering from cerebral palsy were graded according to severity of physical handicap, those with "slight" handicap were found to have a mean intelligence quotient of 81.8, those with moderate handicap 71.7, and severely handicapped patients 55.6. The most severely handicapped group had a mean quotient of only 24.6. This relationship between the severity of physical and intellectual handicaps has been confirmed by most observers, though exceptions occur.

Dunsdon (1952) found that more children with extensive, rather than severe, physical handicaps had low intelligence quotients than had patients with less extensive physical disorders. Approximately 70% of those with all four limbs involved/

involved (whether by paresis or involuntary movement) had quotients of less than 70, whilst 38% of paraplegic and 29% of hemiplegic patients had quotients of less than 70. Four per cent of tetraplegic patients had quotients of 100 or more compared to 13.6% of hemiplegics and 14% of those with paraplegia. These figures were taken from a regional survey and are significantly different from those derived from a selected population of candidates for a school for spastic children.

Rather similar results were obtained in an investigation carried out in the City of Liverpool by Floyer (1954). She found that the mean intelligence quotient of hemiplegic patients was 80, of paraplegic patients 71, and of children with quadriplegia 45. Thirty-three per cent of children with paraplegia were considered to be ineducable and 65% of those with quadriplegia (a category which probably contained some patients suffering from bilateral hemiplegia).

Woods (1957) considered that 20% of 25 paraplegic patients and 50% of 67 patients suffering from "spastic quadriplegia" were ineducable, compared to 11% of hemiplegic children and 19% of those suffering from athetosis. Eight per cent of paraplegic patients and 2% of quadriplegics were considered to be of superior intelligence compared to 9% of those with hemiplegia and 3% with athetosis.

All these studies tend to confirm the finding that in diplegia the severity and extent of the paresis tends to be parallelled by the severity of intellectual impairment. Paraplegic patients are less severely mentally handicapped than are tetraplegic patients. This observation was borne out /

out in the present survey.

In addition to poor intelligence and motor handicaps there are often other educational disabilities in diplegic patients. Diminished span of attention and poor concentration were noted by Dunsdon (1952) and Schonell (1958). Spatial difficulties which impaired the ability to recognise and orient letters and small words and letters when reading and writing were described by Meyer (1950) and Dunsdon (1952). Personality disturbances, some functional, others apparently of "organic type" resulting in disorganisation of behaviour similar to that seen in hemiplegic patients, have been noted by Bakwin and Bakwin (1951) and Bender (1955). Disorders of body image, though less accessible to study than those observed in more intelligent hemiplegic children have been investigated by Albitrecchia (1958). She stresses the severe effect that such disorders perceptions of "self" have upon the child's whole relationship with his environment.

Speech Disorders.

The prevalence of speech disorders in diplegia has been estimated by a number of authors. Their estimates vary according to their classifications of cerebral palsy and according to what they consider is a significant speech defect. Dunsdon (1952) found speech defects among 39% of paraplegic patients and 71% of children with "spastic quadriplegia" compared to 71% of "athetoid" patients, but did not distinguish between dysarthric and functional disorders.

Asher and Schonell (1950) found speech defects or inability to speak in 52% of patients suffering from "spastic quadriplegia". They /

They stated that "hemiplegics and paraplegics rarely had speech defects." Skatvedt (1958) noted that speech disorders in cerebral palsy might be due "partly to low intelligence, partly to dysco-ordination of the muscles of respiration and speech, partly to different forms of aphasia or reduced hearing. Dysarthria is a very frequent complication in cerebral palsy." She found "dysarthria" as distinct from retardation of speech development in 18.8% of 90 patients suffering from "bilateral spasticity" who were examined for dysarthria. This compared with a prevalence of 10.5% in hemiplegia and 66% in "athetosis". Speech defects in diplegia due to hearing defects are seldom noted by recent authors.

CHAPTER 7a

Family History in 79 Patients Suffering from Diplegia.

PARENTS.

One parent, the father in Case 47, suffered from hemiplegia acquired in childhood and with complicating epilepsy and behaviour abnormalities.

Other neurological abnormalities were present in the parents of three cases. In Case 22 the mother was deaf as a result of otosclerosis and had blue sclerotics. Her elder child, the brother of the patient, suffered from osteogenesis imperfecta. In Cases 62 and 174 the fathers suffered from disseminated sclerosis and severe residual poliomyelitis respectively.

Only one parent of a patient with diplegia (Case 206) suffered from epilepsy which had been present from childhood.

Feeble mindedness was present in one or both parents of 16 patients. All but one of the cases were in Social Classes IV and V, the exception being Case 155 in which the mother only was feeble minded. In 6 cases both parents were feeble-minded; in 8 only the mother and in 2 only the father. In 5 of the 16 cases in which one or both parents were feeble minded, siblings of the patients were similarly affected.

Psychiatric disturbances or alcoholism were present in 11 cases. In 4 cases these affected the father only, the disorder being alcoholism in Case 61, anxiety neurosis in Cases 96 and 103, and psychopathy which had resulted in life imprisonment for murder in Case 53. In 5 cases psychiatric disturbances affected the mother only. In Case 12 the mother had /

TABLE 71

HEALTH OF PARENTS OF DIPLEGIC PATIENTS

A - Neurological Conditions and Epilepsy

Case No.	Maternal Age	Mother	Father	Siblings	Relatives
22	37	Deaf. Otoscler-osis.	Healthy.	1 Fragilitas ossium.	Healthy.
47	40	Anxiety neurosis.	Hemiplegia and epilepsy.	1 mentally retarded. 1 mentally disturbed. 1 normal.	Mother's brother mentally retarded. Mother's sister psychotic.
62	39	Healthy.	Disseminated sclerosis.	1 healthy.	Healthy.
174	23	Anxiety neurosis.	Severe poliomyelitis.	1 epileptic. 1 healthy. 1 tuberculous.	2 brothers of mother congenital malformations.
206	30	Epilepsy.	Healthy.	2 healthy.	Mother's nephew epileptic.

had been treated for obsessional fears, and in Case 174 for anxiety neurosis. In Case 154 she was psychotic and feeble minded. Since in Case 193 the mother greeted the examiner with a threat of legal proceedings, it was difficult to form an accurate clinical impression, but the general practitioner thought she was paranoid, "but I daren't refer her for treatment - you try." The mother in Case 200 was a psychopathic prostitute with a criminal record. In Case 40 the mother suffered from anxiety neurosis after divorcing her husband on account of violence and alcoholism. In Case 177 the mother was psychopathic and divorced from her husband who was an alcoholic Regular Army sergeant.

Physical Health.

In 19 cases one or both parents suffered from physical disorders which were not of neurological origin. In 5 cases physical disease was confined to the father's side (Cases 2, 25, 56, 128 and 154), though the mother was considered to be feeble minded in Cases 25 and 128, and possibly psychotic in Case 154. In Cases 2 and 56 the fathers suffered from duodenal ulcers and were of Social Class III. In the other three they were of Social Classes IV or V. In Case 25 the affected father was feeble minded and had rheumatic mitral stenosis. In Case 128 the father was of rather low intelligence and suffered from chronic bronchitis, resulting in frequent unemployment. In Case 154 the father was an unstable and sometimes alcoholic Pole, at times separated from his wife. He suffered from chronic pulmonary tuberculosis.

In /

In 9 cases the physical health of the father was good, but that of the mother abnormal (Cases 45, 53, 59, 124, 138, 139, 140, 186 and 207). In Case 45 the small mother suffered from pyelitis before and during her pregnancy. In Case 138 the mother found that her chronic bronchitis was exacerbated by pregnancy. In Case 186 the mother was thyrotoxic before her pregnancy which was complicated by pre-eclampsia, and there was only slight remission during it. Three mothers were menopausal at the time they conceived the affected child (Cases 53, 59 and 207). In Case 53 periods had been scanty and irregular for some time and the mother had become increasingly obese and bronchitic. In Case 59 also the mother, who was feeble minded, was obese. In Case 207 the mother refused to go into details but said she "had good reasons for thinking she was past it" at the time she conceived the patient. In Cases 124 and 139 retroversion of the uterus was treated prior to conception. In Case 124 a pessary was inserted following dilatation and curettage of the uterus, and in Case 139 a sling operation was performed 2 years before conception. In Case 140 left ovariectomy had been performed two years before conception, when the abdomen was explored on account of a feeling of heaviness and pelvic pain.

In 5 cases both parents had poor physical health (Cases 10, 13, 63, 157 and 161). In Case 13 both parents suffered from pulmonary tuberculosis and the mother had had an ovariectomy performed the year before she became pregnant at the same time as a perforated appendix was removed. In Case 63 the /

the mother suffered from chronic pulmonary tuberculosis said not to be active at the time of her pregnancy, and the father was a paraplegic as a result of war wounds. In Case 10 the mother suffered from nephrolithiasis with recurrent urinary infections, and the father from chronic bronchitis. In Case 157 the mother was thyrotoxic and also suffered from iron resistant anaemia which was severe during her pregnancy. The father suffered from cholelithiasis and a duodenal ulcer, both of which had been operated upon with little apparent success. In Case 161 the mother was deaf as a result of chronic otitis media with profuse bilateral stinking discharge, and the father had tuberculosis of the hip which had remained active since first noted at the age of about 14 years.

The frequency of ill-health in the parents.

A consideration of the prevalence of feeble mindedness, frank mental defect, psychiatric disorders and physical disease amongst the parents of diplegic patients shows that a remarkably high percentage were physically or mentally abnormal (Table 71).

In Table 71. it is shown that there was feeble mindedness in one or both parents in 17 families (22%), and psychiatric disturbance in one or both in 12 (16%). Physical disease was present in one or both parents in 25%. In a total of 58% of the families one or both parents was unhealthy, and neither parent was healthy in as many as 20 of the 75 families (27%).

SIBLINGS. /

SIBLINGS.

Cerebral palsy was present in one sibling in each of 3 cases, of which two consisted of a twin pair. In Cases 158 and 159 twins were affected, the clinical findings being very similar in both of them. One had accessory auricles and the other a congenital heart lesion. In Case 98 the younger brother of the patient suffered from hemiplegia of unknown origin (Case 199). A further sibling and two siblings of the mother were feeble minded.

Epilepsy was present in a sibling in 3 cases (Cases 103, 128 and 174). In the latter the mother's brother was also epileptic, and in Case 174 the mother was feeble minded.

Mental retardation was present in one or more siblings in 10 cases (Table 72). Two siblings were affected in Cases 41, 42 and 118. Other relatives were feeble minded or mentally defective in three cases (Cases 98, 114 and 177). Of the 10 cases in which siblings were mentally retarded, one or both parents were similarly affected in 5.

In Case 128 one of the mentally defective siblings also suffered from congenital heart disease. In Case 22 one of the siblings suffered from osteogenesis imperfecta. In Case 28 a sibling had died after the age of one year with multiple congenital abnormalities. Death of siblings during infancy was attributed to spina bifida in Case 41, multiple congenital abnormalities in Case 42, convulsions in Cases 174, 205 and 76, and two siblings in Case 157. It seems probable that some of the children who died with convulsions may have had congenital anomalies /

TABLE 72

DIPLEGIA - HEALTH OF SIBLINGS

A. Neurological abnormalities and mental deficiency

Cerebral Palsy

Case No.	Age of mother	Mother	Father	Healthy siblings	Abnormal siblings	Other relatives
98	33	Feeble-minded.	Feeble-minded.	2	1 brother hemiplegic. 1 sister mentally retarded.	None known abnormal.
158 159 twins	23	Feeble-minded.	Healthy	0	Both diplegic.	None known abnormal.

Epilepsy

128	33	Poor intelligence. and anxious.	Poor intelligence. Chronic bronchitis.	3	One brother feeble-minded. Congen. heart disease. One brother mentally defective and epileptic.	None known abnormal.
174	23	Anxiety neurosis.	Severe poliomyelitis.	2	1 sister epileptic. 1 sister tuberculous.	None.
103		Healthy.	? Psychotic	0	Patient's half-sib. (by father) epileptic.	Mother's brother epileptic.

Mental retardation

47	Anxiety neurosis.	Hemiplegia and epilepsy.	1	1 Psychotic. 1 Mentally defective.	Mother's brother mentally defective.
98	Mentally retarded.	Mentally retarded.	1	1 Hemiplegic. 1 feeble-minded.	1 brother and sister of mother mentally retarded.
114	Feeble-minded.	Healthy.	1	1 Feeble-minded.	Father's sister mentally defective.
[illegible]	[illegible]	[illegible]	2	1 Feeble-minded.	Mother has diplegic cousin.
76	Healthy.	Healthy.	1	1 Mentally defective.	
157	Thyrotoxic. Anaemic.	Gall-stones. Duodenal ulcer.	1	1 Mentally defective.	
128	Feeble-minded	Chronic bronchitis.	5	1 Mentally defective 1 Mentally defective and congenital heart.	

TABLE 72

DIPLEGIA - HEALTH OF SIBLINGS

B. Other abnormalities

Case No.	Age of mother	Mother	Father	Healthy siblings	Abnormal siblings	Other relatives
22	37	Blue schlerotic otosclerosis.	Healthy	0	1 brother osteogenesis imperfecta.	Healthy.
174	23	Anxiety neurosis.	Severe poliomyelitis.	2	1 Epileptic sister. 1 Tuberculous.	Healthy.
128	33	Poor intelligence.	Chronic bronchitis.	3	1 brother feeble-minded and congenital heart disease. 1 Mentally defective and epileptic.	Healthy.

anomalies of the nervous system.

Thus, of the total of 96 full siblings, one had a hemiplegia probably of congenital origin, two were epileptic, one had congenital heart disease, and one osteogenesis imperfecta. Including one of the cases of epilepsy and the case with congenital heart disease, 14 were mentally defective or feeble minded - approximately 15%. In addition one was thought to be psychotic (Case 47) and was of poor intelligence. Thus a total of 18 full siblings were abnormal - approximately 19%.

When the distribution of the abnormal siblings is considered it is interesting to note that there were 4 cases with more than one abnormal sibling (Cases 41, 42, 47 and 98). In 3 cases both parents were feeble minded and produced between them 9 abnormal siblings, 4 healthy siblings, 7 abortions, and 5 neonatal deaths. In Case 47 the father had hemiplegia and epilepsy and the mother anxiety neurosis. Including the patients, the 4 mothers had a total of 30 pregnancies, only 5 of which resulted in healthy children.

When the 12 cases with abnormal siblings are considered, it is noted that one or both parents were physically abnormal in 6 cases, mentally subnormal in 5 cases and suffering from psychiatric disorder in three. Both parents were healthy in one case (8%), one parent was normal in two cases (17%), and both were abnormal in 9 cases (75%). In these 12 cases, 20 of the 24 parents were abnormal (84%), which is a proportion significantly different statistically from that which might be expected amongst all diplegic parents. The increased

proportion /

proportion of cases in which both parents were abnormal is also significantly different from that which would be expected on the basis of chance alone. Another way of looking at this association (of two abnormal parents having a diplegic child and also abnormal siblings) is that of the 20 patients both of whose parents were abnormal, 9 also had siblings who were abnormal.

OTHER RELATIVES.

A history of other relatives suffering from cerebral palsy was elicited in 3 cases of diplegia. In Case 10 the patient's cousin, the son of the mother's sister, was diplegic. The maternal grandmother and another of her daughters were feeble minded. In Case 177 the mother's cousin by the maternal grandmother's sister was diplegic. A sibling of the patient was feeble minded. In Case 163 a cousin of the father by the paternal grandmother's sister was diplegic.

A history of epilepsy amongst the relatives of patients with diplegia was elicited in 4 cases. The mother's nephew, the son of her sister, was affected in Case 206, and the mother herself suffered from recurrent convulsions. In Case 12 the maternal grandfather and uncle were epileptic. In Case 40 the father's brother died of convulsions, and his nephew, son of the affected brother, died of a cerebral glioma, a complication of generalised neurofibromatosis. In Case 103 the mother's brother was epileptic, as was the patient's half-sibling.

A /

TABLE 73

History of Abnormal Relatives of Diplegic Patients

Case No.	Abnormality in relative	Health of Mother	Health of Father	Health of siblings	Other normal pregnancies and labours	Abnormal pregnancies and labour	Abortions & Stillbirth Neonatal and Infant Deaths
10	Mother's sister and aunt feeble-minded. Cousin of patient diplegic.	Nephrolithiasis	Chronic bronchitis	1 healthy	1	0	None
12	Maternal grandfather and uncle epileptic.	Anxiety neurosis.	Healthy	1 healthy	0	1	None
31	Maternal grandfather psychotic.	Feeble-minded.	Healthy	1 healthy	0	1	1 abortion
40	Father's brother died with epilepsy. Nephew died cerebral tumour, Neurofribromatosis.	Anxiety neurosis.	Psychopathic Alcoholic	2 healthy	2	0	None
47	Mother's brother mentally defective and sister psychotic.	Anxiety neurosis.	Hemiplegia and epilepsy.	1 Mentally def. 1 ? psychotic 1 normal	1	2	None
98	One brother and sister of mother mentally defective. Some defective on father's side.	Feeble-minded.	Feeble-minded.	1 Hemiplegic. 1 feeble-minded. 1 well.	0	1	2 abortions
103	Mother's brother epileptic, Patient's half-brother (by father) epileptic.	Healthy.	Very unstable. ? Psychotic.	Patient's half-brother by father mentally defect.	0	0	None
114	Father's sister mentally defective.	Feeble-minded.	Healthy.	1 Feeble-minded. 1 Healthy.	0	2	None
118	Maternal grandparents deaf and dumb.	Healthy.	Healthy.	1 Healthy.	1	0	1 previous abortion
163	Cousin of father severely mentally defective and may be diplegic.	Healthy.	Healthy.	2 Healthy.	2	0	None
174	2 brothers of mother died of multiple deformities (? spina bifida) in infancy.	Anxiety neurosis.	Severe polio-myelitis.	1 Epileptic. 1 Healthy. 1 Tuberculous.	0	4	One neonatal death
177	Mother has diplegic male cousin.	Psychopathic.	Alcoholic.	1 Feeble-minded. 2 Healthy step-brothers.	1	2	None
206	Nephew of mother epileptic.	Epileptic.	Healthy.	2 Healthy.	2	0	None

A history of mental retardation amongst relatives was elicited in 5 cases. In Case 10 the mother's sister and aunt were feeble minded, as her nephew, the son of another of her sisters, was diplegic. In Cases 47 and 98 siblings of the mother were mentally retarded. In Case 114 the father's sister was mentally defective, and in Case 163 the father's cousin. In two of these cases one or both parents and in 3 cases one sibling was feeble minded.

In Case 118 both maternal grandparents were deaf mutes. In Case 174 two of the mother's brothers died of multiple congenital abnormalities including spina bifida in infancy. Psychosis was present in the maternal grandfather in Case 31 and in the mother's sister in Case 47, in which the father himself was hemiplegic and suffered from epilepsy.

Consanguinity.

The parents were certainly related in two cases; in Case 76 they were first cousins born to sisters of the same grandparents, and in Case 133 they shared great-grandparents. In Case 76 both parents were healthy and they would admit no other family abnormality (though they were extremely reticent altogether). Their first child was mentally defective, their second was healthy. Their third died at the age of 6 months with convulsions attributed to diphtheria, and their fourth was the feeble minded diplegic patient.

In Case 133 the parents came from Lewis and were healthy. They refused information about any other members of the family, and the doctor who was written to in order to obtain more details /

details turned out to be a distant relative. In addition to the patient they had one healthy child.

The parents came from the same locality and there was a possibility of distant relationship in Case 47, in which both came from Shetland, and both had unstable and defective relatives (Table 74).

Consideration of the family histories of diplegic patients.

Since the total population of relatives is unknown, the prevalence of cerebral palsy amongst them cannot be calculated. The fact that all the more distant relatives affected were cousins either of the patients or one of his parents and that they all suffered from diplegia suggests a Mendelian recessive mode of inheritance.(Table 74). The presence of consanguinity of the parents in at least two and probably three cases would further favour this hypothesis.

Since the survey was completed further study has been made of the possibility that a proportion of cases of diplegia are the result of Mendelian recessive inheritance, as suggested by Penrose (1938). Further cases are being studied and it seems certain that a minority are the result of transmission by a Mendelian recessive gene. One difficulty encountered in determining the importance of genetic factors in diplegia has been that the families of mothers whose children suffer from diplegia tend to be small. It is probable also that a relatively high proportion of affected children perish as a result of stillbirth or neonatal death before the diagnosis can be established. It is hoped that further research which is /

TABLE 74

Family History of Cerebral Palsy amongst Diplegic Patients

Case No.	Mother	Father	Healthy siblings	Abnormal siblings	Stillbirths or infant deaths.	Relatives
10	Nephrolithiasia	Chronic bronchitis	1	-	-	Mother's sister and aunt feeble-minded. Patient's cousin by mother's sister diplegic.
177	Psychopathic.	Alcoholic.	2	1 Feeble-minded.	-	Mother's cousin diplegic.
163	Healthy.	Healthy.	2	-	-	Father's cousin diplegic.
47	Healthy.	Hemiplegia and epilepsy.	1	1 Mentally defective. 1 ? psychotic.	-	Mother's brother mentally defective. Mother's sister psychotic.
98	Feeble-minded.	Feeble-minded.	1	1 Hemiplegic. (Case 199)	-	One brother and one sister of mother mentally defective.
158 159	Twins. Feeble-minded.	Healthy.	0	-	-	-

is being actively pursued will help to elucidate the importance of Mendelian recessive inheritance in diplegia.

As in hemiplegia, the significance of the prevalence of feeble mindedness amongst siblings and parents may be interpreted in a number of different ways. Penrose suggested that true microcephaly might be another manifestation of the gene responsible for diplegia. Unfortunately the heads of the siblings of patients in the present series were not measured. Since the prevalence of mental retardation amongst parents (approximately 14%) was almost the same as that amongst siblings, there is little evidence that the mental retardation observed was inherited as a Mendelian recessive trait (Table 75). The extent to which mentally retarded parents were predisposed to produce diplegic children because of genetic factors and how much by environmental conditions is extremely difficult to assess.

The poor physical health of parents and especially of mothers is striking. In particular a high proportion of mothers were menopausal or had suffered from gynaecological disorders, which might be expected to impair their reproductive efficiency. Without a control series of normal children with which to compare the abnormalities of mothers it is difficult to assess the significance of this finding. The fertility of mothers of diplegic patients will be discussed later.

The prevalence of epilepsy amongst parents, siblings and relatives is probably not significantly greater than that in the general population (Table 76).

TABLE 75

Family History of Mental Retardation

Case No.	Diagnosis of Patient.	Mother	Father	Healthy siblings	Abnormal siblings	Relatives
10	Diplegia.	Nephrolithiasis.	Chronic bronchitis.	1	-	Father's sister mentally defective. Mother's sister and aunt feeble-minded. Cousin of patient diplegic.
47	Diplegia.	Anxiety neurosis.	Hemiplegia and epilepsy.	1	1 "Psychotic" 1 Mentally defective.	Mother's brother mentally defective.
98	Diplegia.	Mentally retarded.	Mentally retarded.	1	1 Hemiplegic. 1 Feeble-minded.	One brother and one sister of mother mentally defective.
114	Diplegia.	Feeble-minded.	Healthy.	1	1 Feeble-minded.	Father's sister mentally defective.
163	Diplegia.	Healthy.	Healthy.	2	-	Father's cousin severe mental defect. Probably diplegic.
177	Diplegia.	Psychopathic.	Alcoholic.	2	1 Feeble-minded.	Mother has diplegic cousin.
41	Diplegia.	Feeble-minded.	Feeble-minded.	5	2 Mentally defective.	-
42	Diplegia.	Feeble-minded.	Tuberculous. Feeble-minded.	0	1 Mentally defective. 1 Feeble-minded.	-
76	Diplegia.	Healthy.	Healthy.	1	1 Mentally defective.	-
157	Diplegia.	Thyrotoxic. Anaemia.	Gallstones. Duodenal ulcer.	1	1 Mentally defective.	-
25	Diplegia.	Feeble-minded.	Feeble-minded. Rheumatic heart disease.	3	-	-
31	Diplegia.	Feeble-minded. Obese.	Healthy.	1	-	-
38	Diplegia.	Healthy.	Feeble-minded. Died.	3	-	-
59	Diplegia.	Feeble-minded. Obese. Menopausal.	Feeble-minded.	1	-	-
75	Diplegia.	Feeble-minded.	Healthy.	2	-	-
128	Diplegia.	Feeble-minded.	Chronic bronchitis.	5	1 Mentally defective. 1 Mental defect and congenital heart.	-
131	Diplegia.	Feeble-minded. Epileptic.	Feeble-minded.	0	-	-
154	Diplegia.	Psychotic. Feeble-minded.	Tuberculous.	1	-	-
155	Diplegia.	Feeble-minded.	Healthy.	3	-	-
156	Diplegia.	Feeble-minded.	Feeble-minded.	1	-	-
183	Diplegia.	Menopausal.	Feeble-minded.	0	-	-
205	Diplegia.	Feeble-minded.	-	6	-	-

TABLE 76

Family History of Epilepsy and Diplegia

Case No.	Mother	Father	Healthy Siblings		Stillbirths or neonatal deaths	Relatives
128	Healthy.	Chronic bronchitis.	3	1 brother feeble-minded, mentally defective and epileptic.	-	-
174	Feeble-minded.	Healthy.	2	1 Tuberculous. 1 sister epileptic.	1	-
206	Epilepsy.	Healthy.	2	-	-	Mother's nephew epileptic.
12	Healthy.	Healthy.	1	-	-	Maternal grandfather and uncle epileptic.
40	Healthy.	Psychopathic. Alcoholic.	2	-	-	Father's brother died epilepsy. Father's nephew died neuro-fibromatosis.
103	Healthy ?	Psychotic	0	Patient's half-brother (by father) epileptic	0	Mother's brother epileptic.

Maternal Factors in Diplegia.

The physical and mental health of the mothers of diplegic patients has been described, but the physical disorders merit further consideration from the point of view of the possible effects on the ability of the mothers to childbear successfully.

It will be seen from Table 77 that, excluding purely neurological, mental and psychiatric disorders, a total of 14 mothers was thought to be unhealthy. In 7 chronic infections were present, recurrent urinary infections with nephrolithiasis in Case 10, recurrent pyelitis in Case 45, bronchitis in Cases 53 and 138, very severe chronic otitis media in Case 161 and tuberculosis in Cases 13 and 63.

The mothers of 3 patients were menopausal at the time of conception (Cases 53, 59 and 207). All were obese and Case 53 also suffered from chronic bronchitis. All three cases were 42 years old. All had had previous pregnancies.

Four mothers had gynaecological disorders. In Case 124 the mother had a retroverted uterus which had to be corrected during pregnancy. In Case 139 retroversion had previously been corrected by a sling operation. In Cases 13 and 140 there had been previous ovariectomy, in Case 13 on account of an ovary being involved in an appendicial mass, and in Case 140 on account of abdominal pain occurring during pregnancy.

Two mothers were thyrotoxic before and during pregnancy (Cases 157 and 186). In the former there was associated anaemia, and pregnancy was complicated in both by pre-eclampsia, labour being premature. In at least 6 other mothers there was a history of very irregular menstruation from the time of puberty, but most unfortunately questioning about the time of /

TABLE 77

Health of Mothers of Diplegic Patients

Case No.	Age	Mother's health	Father's health	Other pregnancies	Siblings	Relatives	Birth weight
10	38	Nephrolithiasis Recurr.urin. infect.	Chronic bronchitis.	One normal	1 healthy.	Mother's sister and aunt feeble-minded. Cousin of patient diplegic.	9/8
13	22	Tuberculosis. Ovariotomy.	Feeble-minded.	1 forceps	1 healthy.	Healthy.	3/6½
45	23	Very small 5'0" Pyelitis.	Healthy.	None	None	Healthy.	4/7
53	42	Obese. Bronchitis. Menopausal.	Psychotic. In gaol.	2 prem.deliv. 1 normal.	3 healthy.	Healthy.	2/8
59	42	Feeble-minded. Obese. Menopausal.	Feeble-minded.	1 healthy.	2 healthy includ.twin.	Healthy.	5/13½
63	31	Old tuberculosis.	War diabetic.	1 premature.	1 healthy.	Healthy.	3/13
124	37	Retroverted uterus. Otherwise well.	Healthy.	1 healthy.	1 healthy.	Healthy.	3/8
138	23	Chronic bronchitis.	Healthy.	None	-	Healthy.	8/5½
139	31	Retrover. of uterus.	Healthy.	2 normal.	2 healthy.	Healthy.	6/13
140	33	Ovariotomy.	Healthy.	2 normal. 1 prem. onset of labour.	3 healthy.	Healthy.	3/10½
157	34	Thyrotoxic. Anaemic.	Gall-stones duodenal ulcer.	4 normal	2 inf. deaths. 1 healthy. 1 ment. deft.	Healthy.	8/0
161	24	Deaf.Chronic otitis media.	T.B. hip.	2 healthy.	1 Inf. death. 1 normal.	Healthy.	7/5½
186	34	Thyrotoxic.	Healthy.	1 normal.	1 healthy.	Healthy.	5/2¾
207	42	Menopausal. obese.	Healthy.	1 normal.	1 healthy.	Healthy.	7/8

TABLE 78

Abnormal menstruation in mothers of diplegic patients

Case No.	Total number of pregnancies	Number of abortions, stillbirths and neonatal deaths	Pregnancy and delivery
Irregular			
31	1/3	1 abortion	Antepartum haemorrhage. Premature delivery.
81	1/2	-	External version. Transverse arrest. Prolonged labour. Mature.
Scanty			
34	4/4	1 stillbirth	Mature. Medical induction. Precipitate labour.
53	4/4	-	Precipitate premature delivery.
59	2/2	-	Mild pre-eclampsia. Normal mature delivery.
183	1/1	-	Normal premature delivery.
Late onset			
103	1/1	-	Uncomplicated spontaneous mature delivery.
139	1/3	-	Uncomplicated spontaneous mature delivery.

of onset of menstruation, its regularity and its duration was not made routine until towards the latter part of the survey.

It is of interest that as many as 7 of the 14 mothers with physical disease should be either menopausal or have disorders of the reproductive organs (Table 78). Clearly the significance of these disorders, like that of thyrotoxicosis in another two cases is extremely difficult to assess without large control series of cases. Nevertheless, there is a suggestion that disturbances of endocrine balance during pregnancy may be of importance directly or indirectly in the aetiology of diplegia.

It is also difficult to attempt to assess the significance of the presence of chronic infections in the mothers. It is possible that both maternal and foetal nutrition suffered as a result of them.

Maternal Age.

The distribution of diplegic patients by the age of their mothers is indicated in Table 80 where it is compared to that for all live births in Scotland shown by the Registrar General (1951). It will be seen that there is a higher proportion of mothers over the age of thirty in the diplegic series. This is in spite of the fact that there is a greater proportion of first-born diplegic children than there are first-born children in the community. The difference is not statistically significant, however, when the figures are presented in their present form. As the table indicates, the ages of mothers with diplegic patients are very similar to those /

those of mothers bearing children with congenital hemiplegia, and older than those with children suffering from acquired hemiplegia. The average maternal age is about exactly the same as that found by Vannet (1944) in 52 "diplegic" patients, some of whom were probably not diplegic.

Table 79

Average Age of Mothers at the Time of the Birth of their Diplegic Child

	Average Age
1st and only child	27
1st of number of children	26
2nd child	33
3rd child	31
4th and subsequent	33
All patients	29

Table 80

The Distribution of 75 Diplegic Patients by Maternal Age at the Time of Birth Compared to the Distribution of all Scottish Live Births (1951)

Maternal Age	No. of Cases	Approx. %	Registrar General %
15 years but under 20	7	10	4
20 " " " 25	20	26	27
25 " " " 30	12	16	32
30 " " " 35	24	31	22
35 " " " 40	10	13	12
40 and over	3	4	3

Size of Families in Which there is a Diplegic Child.

In 17 cases (23%) of those in whom the family size was known /

known, the diplegic child was the only child. In 30 cases (approx. 40%) there was one sibling; in 13 cases (approx. 17%) there were two siblings, and in 9 (approx. 12%) there were 3 siblings. In 6 (approx. 8%) there were 4 or more siblings. Thus at the time of the examination each mother of a diplegic child had an average of 2.5 children including the patient. This compares with an average of 2.8 per mother for congenital hemiplegia and 3.6 for acquired hemiplegia.

Place in Family.

The place in the family of the diplegic patient is shown in Table 79 . It will be seen that there is a statistically significant preponderance of first-born diplegic children ($p > 0.01$) compared to that for all live births in Counties of Cities (Registrar General for Scotland, 1951). This is in apparent agreement with the majority of authors who emphasise the high proportion of first-born children who have diplegia. (Table 81)

Unfortunately, however, this is not a valid observation for it takes no account of family size. When the distribution by place in the family is compared with the expected chance distribution (Table 82) it is found that there is no statistically significant difference between them. The observed and expected distributions are, in fact, remarkably similar, except for a lower proportion of first-borns in the observed group.

The frequently published observation that first-born children are more likely to show diplegia than second or third-born children is not confirmed, though it is true that a /

TABLE 82

Number of surviving children in the families of patients with diplegia, in which family size known

	Number of cases	% of cases
Patient the only child. *	17	23
Patient and one sibling.	30	40
Patient and two siblings.	13	17
Patient and three siblings.	9	12
Patient and four or more siblings.	6	8
	75	100

* including the twins, Cases 158 and 159 as one not two cases, for this purpose.

TABLE 81

The distribution of diplegic patients by place in family to that of all live births in Scottish Counties of Cities (Registrar General, 1951)

	Diplegia.		Approximate %		Counties of Cities approx. %
First and only.	17	38	22	50	35
First of two or more.	21		28		
Second.	20		27		29
Third.	8		10		16
Fourth.	7		9		9
Fifth and over.	3		4		11
Unknown	2		-		-
Total	78		100		100

a high proportion of diplegic patients will be first-born as the average size of the families of these patients is small. A further indication of this is found in the fact that the average number of previous live and stillborn siblings in the case of each diplegic child is 1.0. For the general population it is about 1.8, and for mothers of patients with acquired hemiplegia it is 1.9. Excluding those cases in which the diplegic child was the first child, the average number of previous live and stillbirths was 1.9, compared to 2.4 per multiparous mother of a child with congenital hemiplegia and 3.0 per mother with a child suffering from acquired hemiplegia.

The Spacing of Pregnancies.

Since the high proportion of miscarriages occurring at about the time of the pregnancy with the diplegic patient suggested that there might be some temporary disturbance of the mother's ability to carry children at these times, other indications of this were sought.

It seemed possible that if there were diminished fertility at about the time of the pregnancies with the patients, then fewer children might be born immediately before and after the patients than at other times. This would be reflected in a greater lapse of time between the birth of the immediate predecessors and successors of the patients than amongst the other children. A study was therefore made of the lapse of time between the birth of the patients and their siblings, and also the occurrence of miscarriages. The significant

results /

results of this are shown in Table 83 .

It will be seen that the average lapse of time between the births of non-diplegic siblings delivered before the birth of the patients was 1.8 years, and of those born after the diplegic child 3 years. The average lapse of time between the births of all non-diplegic siblings was almost exactly two years.

On the other hand there was an average period of 4.2 years between the birth of the immediately preceding sibling and the diplegic child, and if abortions are included an average lapse of 3.8 years between previous delivery or abortion and the birth of the patient. The average period between the birth of the diplegic child and the birth of the succeeding child was 3.6 years.

No doubt contraception may have played a part in reducing the number of children conceived after the birth of a diplegic child. But the greater time that elapses between the birth of the preceding child and the conception of the diplegic patient than between other siblings strongly suggests that there must be diminished maternal fertility at about the time the diplegic child is conceived.

A study of the other pregnancies of mothers of diplegic children.

Including the pregnancies resulting in the birth of the diplegic patients (three of which were twin) there were 214 pregnancies - 197 still and live born infants and 20 abortions. Of the 197 still and live-born children, 76 were diplegic (including twins), and there were healthy twin siblings of the patients in two other cases (Cases 41 and 63).

Eighty-eight /

TABLE 83

Average period of time between the births of siblings and of diplegic patients and their siblings

1.	Average period between the births of siblings. (96 contiguous siblings)	2 years.
(a)	Average period between the births of siblings prior to the birth of the diplegic patient. (68 contiguous siblings).	1.8 years.
(b)	Average period between the births of subsequent siblings. (28 contiguous siblings).	3.0 years.
2.	Average period between the birth of the diplegic child and either prior or subsequent sibling. (64 siblings).	4.0 years.
(a)	Average period between birth of preceding child and patient. (40 cases).	4.2 years.
(b)	Average period between the birth of the diplegic child and subsequent sibling. (24 cases).	3.6 years.
3.	Average period between last delivery or abortion and the diplegic pregnancy. (42 cases)	3.75 years.

Eighty-eight pregnancies occurred before the birth of the diplegic child and 17 of them terminated in abortion. Fifty-one pregnancies occurred after the birth of the patients and 3 of these ended in abortion. Of the 71 previous pregnancies which did not end in abortion, one ended in stillbirth as a result of haemorrhage from a placenta praevia, one was complicated by pre-eclampsia and two by antepartum haemorrhage. Precipitate delivery occurred in 3 cases, prematurity was the only abnormality in two, labour was prolonged in two and forcpes were used in 7. Only 49 (approx. 56%) of the 88 pregnancies and deliveries occurred entirely without complications. Of 70 live-born infants resulting from these pregnancies, 10 had succumbed by the age of one year. The total number of one-year-old survivors was 60 out of 88 pregnancies.

Of the 51 pregnancies which occurred after the delivery of the diplegic patient, 3 ended in abortion and 2 in premature delivery. Antepartum haemorrhage complicated pregnancy in 2 cases and precipitate delivery in another two. Forceps delivery was required in one case and labour was prolonged in four. Thirty-six (approx. 61%) of the 51 subsequent pregnancies were normal throughout pregnancy and delivery.

Thus of the total of 139 pregnancies, to 59 mothers who had pregnancies additional to those resulting in the diplegic child, only 85 (approximately 61%) were uncomplicated and resulted in the spontaneous vertex delivery of an infant at term. Of the 59 mothers with other pregnancies, only 32 (53%) /

TABLE 84

Comparison of the proportion of abnormal pregnancies and deliveries amongst prior and subsequent pregnancies to mothers giving birth to mature and premature diplegic patients.

	Mature Group.	Premature Group	All
Number of previous pregnancies and deliveries	41	47	88
Number of abnormal previous pregnancies and deliveries.	14	25	39
% of abnormal previous pregnancies.	34	53	44
Number of subsequent pregnancies and labours.	29	22	51
Number of subsequent abnormal pregnancies and labours.	3	12	15
% abnormal subsequent pregnancies and labours.	13	55	34
Total of other pregnancies.	70	69	139
Total of other abnormal pregnancies.	17	37	54
% of abnormal subsequent pregnancies and labours.	24	54	39

(53%) had had only normal other pregnancies.

There is an interesting comparison between the obstetric histories of the mothers giving birth to premature diplegic patients and those giving birth to mature diplegic patients (Table 84,85). The percentage of abnormal pregnancies prior to that resulting in the delivery of the affected patient was 34 amongst mothers of mature patients and 53 for mothers of premature patients. Amongst subsequent pregnancies the percentages are 13 and 55, a still more striking difference. When all previous and subsequent pregnancies are considered it is found that 24% of those to mothers in the mature group are abnormal and 55% of those to mothers in the premature group. These differences are statistically significant. A similar though less marked difference is found in the proportion of abnormal pregnancies and deliveries resulting in the births of the patients between the mothers in the mature and those in the premature groups, as will be described in detail later. Whereas approximately 85% of the 34 pregnancies and deliveries in the premature group were abnormal, in the mature group only about 64% of 41 were abnormal, a statistically significant difference.

Though the higher proportion of pregnancies resulting in abortion amongst the mothers in the premature group compared to the mature group is statistically significant, the differences in the number of neonatal deaths, infant deaths and abnormal siblings between the groups are not.

It is apparent from these figures that there is a relatively high incidence of abnormality in other pregnancies compared /

TABLE 85

Comparison of the numbers of abnormal pregnancies, abortions, stillbirths, neonatal deaths, infant deaths and abnormal children born to mothers of mature diplegic patients and to mothers of premature diplegic children.

	No. of mature	Approx. %	No. of Premature	Approx. %
Number of mothers.	41		34	
Number of other pregnancies.	70	100	69	100
Number of other normal pregnancies.	53	75.7	32	45
Number of other abnormal pregnancies.	17	24.3	37	55
Number ending in abortion.	3	4.3	17	24.5
Number ending in premature birth.	1	1.4	4	6
Number resulting in stillbirth.	1	1.4	0	0
Number ending in neonatal death.	2	2.8	4	6
Number ending in infant death.	2	2.8	2	3
Number of offspring mentally retarded.	6)	8.4)	7)	10.5)
Number of offspring with physical anomalies.	9) 12.6 3)	) 4.2)	9) 13.5 2)	) 3)

compared with that which gave birth to the patient amongst mothers of diplegic children. A more accurate idea of the obstetric histories of these women may be obtained, however, by considering the pregnancy which produced the patient as well as the other pregnancies of the mother (Table 86). It will be seen from this table that only 100 of the total of 214 pregnancies to 75 mothers (47%) were uncomplicated during either pregnancy or delivery. In 53% of pregnancies pregnancy itself or the mode of delivery was abnormal.

Table 86

	Number	%
Total number of pregnancies including those resulting in the birth of patients.	214	100
Number of pregnancies and deliveries which were normal.	100	47
Number in which pregnancy was disordered or delivery abnormal.	114	53

Abortions to Mothers of Diplegic Patients.

Twenty of the 138 other pregnancies to the 75 mothers of diplegic patients about whom details are known ended in abortion (about 14%). The abortions occurred in 13 mothers, 3 of whom had two (Cases 98 and 152) and one of whom had five (Case 41) (Table 87). An interesting point about the timing of the abortions was that 13 of them occurred immediately prior to the diplegic pregnancy or to another abortion preceding the diplegic pregnancy - a significantly high proportion statistically. Thirteen of the 20 abortions occurred within two years before or two years after the birth of /

of the diplegic patient.

In spite of questioning it was impossible to obtain any reliable information about the numbers of abortions which were procured. At least two (Cases 41 and 189) are thought to have been.

None of the mothers with abortions was considered to have poor general health. Four were considered to be of subnormal intelligence (Cases 31, 41, 75 and 98) and none to suffer from psychiatric disturbances. Only one of them had neonatal or infant deaths (Case 41), but three had mentally subnormal siblings (Cases 41, 47 and 98) and one (Case 98) had a hemiplegic child.

It is of interest that 14 of the 25 other pregnancies to the mothers of patients with diplegia who had abortions were complicated in pregnancy, labour or delivery. Nine of the 14 (63%) were delivered prematurely after complicated pregnancies, labours or deliveries. This high incidence of premature delivery in other pregnancies than those resulting in the births of patients or in abortion is unlikely to be due to chance. The social class distribution of mothers with abortion did not differ significantly from mothers without.

Stillbirths, Neonatal Deaths and Postneonatal Deaths.

Stillbirths and neonatal deaths are shown in Table 88 . One patient had a sibling born dead (Case 34) in which the cause was placenta praevia and antepartum haemorrhage. Two other elder sisters were healthy though both were born precipitously.

There /

TABLE 87

Family and obstetric history of thirteen cases with abortion.

Case No.	Number of aborted preg.	No. of preg. with patient	Total No. of preg.	No. of preg. excluding patient and abortions. Abnormal	Normal	No. of healthy siblings.	No. of unhealthy siblings.	No. of neonatal and infant deaths.
2	2	3	3	1	0	1	0	0
31	3	2	3	0	1	1	0	0
32	3	2	3	1	0	1	0	0
41	4,6,8,10,11	12	14	? 4	? 5	3	4	3
47	4	5	5	1	2	1	2	0
63	2,3	4	4	1	0	2	0	0
75	3	4	4	2	0	2	0	0
98	2,6	1	6	2	1	1	2	0
118	1	2	3	1	0	1	0	0
152	2,3	4	4	0	1	1	0	0
172	1	2	2	0	0	0	0	0
181	2	3	3	1	0	1	0	0
189	1	3	3	0	1	1	0	0

There were four patients with siblings who had died in the neonatal period and it was of interest that three of them (Cases 41, 42 and 174) had surviving abnormal siblings, as has been described. In Case 41 there were three neonatal deaths prior to the birth of the patient, as well as five abortions. The first neonatal death was due to multiple congenital anomalies, including a spina bifida. The second two were premature twins. The remaining members of this family consisted of three healthy siblings including the twin of the patient, two feeble minded, one mentally defective, and one mentally defective with cardiac anomalies. In Case 42 two children born prior to the delivery of the patient had died in the neonatal period as a result of congenital anomalies. The sole remaining sibling was mentally defective, though fortunately thought to be clever by his parents.

In Case 174 there were two healthy siblings, one sister with epilepsy, and one child born before the patient, who died on the seventh day after delivery which had occurred immediately after the onset of an antepartum haemorrhage. The cause of death was given as 'Convulsions, ? Meningitis', but there appeared to be little evidence for the latter.

In Case 205 there were no abnormal siblings, but the first-born child, a girl, died at the age of a week with convulsions following a difficult forcpes delivery.

Postneonatal deaths (those occurring between 1 and 12 months of age) are shown in Table 89 . There were four such in three cases. In Case 176 the parents were healthy, but /

TABLE 89

Post neonatal deaths to mothers of diplegic patients

Case No.	Total number of pregnancies.	Birth rank of patient.	Birth rank of infant deaths.	Abnormality of this pregnancy and labour.	Cause of death.	No. of healthy siblings.	No. of abnormal siblings.
76	4	4	3	None	"Diphtheria" with convulsions aged 6 months.	1	1 mental defect.
157	5	5	1,3	None	Both from bronchopneumonia with convulsions aged 8 mths. and 9 mths. respectively.	0	1 very retarded brother.
161	3	1	3	None	"Encephalitis" following vaccination.	1	0

TABLE 88

Stillbirths and neonatal deaths to mothers of diplegic patients.

Case No.	Total number of preg.	Birth rank of patient.	Birth rank of stillbirths or neonatal deaths.	Health in preg. resulting in stillbirth or neonatal death.	Cause of stillbirth or neonatal death.	Number of surviving normal siblings.	Number of surviving abnormal siblings
Stillbirths							
34	4	4	3	Placenta previa.	Antepartum haemorrhage. from placenta previa.	2	0
Neonatal deaths							
41	14	12 (twins)	1,3 (twins)	1 Unknown 3 Good	1 congen. anomalies includ.spina bifida. 3 Premature twins.	3	4 1 ment.def. 2 feeble-minded. 1 ment.def. heart dis.
42	5	5	2,3	2 Prolonged labour. 3 Forceps delivery.	Both had exomphalos and 3 other abnor.	1	1 1 ment.def. brother.
174	5	3	1	1 Antepartum haemorrhage.	Comvulsions and ? meningitis.	2	1 sister has epilepsy.
205	8	8	1	Forceps delivery.	Convulsions aged 1 week.	6	0

but their first child was a mentally defective boy, their second a healthy boy. Their third died in convulsions at the age of 6 months and was diagnosed (somewhat dubiously) as suffering from diphtheria. The fourth was the patient. In Case 157 the first and third of five children died from "bronchopneumonia" with convulsions within the first year of life. The milestones of both appear to have been retarded. There was one mentally defective sister surviving and one healthy brother.

In Case 161 there was one healthy surviving sibling and the patient. The third of the children died of "encephalitis" about three weeks following vaccination.

Thus there was a total of 12 stillbirths, neonatal deaths and postneonatal deaths to the parents of 8 cases in the series, or 11% of the total of 75 families. Neontal deaths were multiple in two cases and postneonatal deaths in one. It is of interest that in five of the cases in which there were deaths in the first year after birth there were other abnormal siblings additional to the patients. Since in only 12 families of the whole series of 75 studied were there other abnormal siblings, this high proportion with both other abnormal children and neonatal or postneonatal deaths is statistically significant. Similarly the facts that as many as 6 of the 7 cases with neonatal or postneonatal deaths had one or both parents abnormal and that both were abnormal in 5 cases (Cases 41, 42, 157, 161 and 174) are also statistically significant.

On the basis of the information available, however, it is very /

very difficult to assess the number of the siblings who died within the first year of life who may have suffered from congenital anomalies and, in particular, diplegia. The fact that the combined neonatal and postneonatal mortality is over 50 per 1000 live births, about twice the average figure for the country, cannot be explained on the grounds of social class distribution alone. Though other factors may be responsible for it in part it is difficult to resist the conclusion that at least a proportion of those dying had, in fact, congenital anomalies.

If diplegia had been present it is unlikely that it would have been apparent at such an early age. Though it certainly cannot be assumed to have occurred in these siblings the fact that two of the neonatal deaths with convulsions, and one from multiple deformities including spina bifida, and that all four of the postneonatal deaths were with convulsions, certainly suggests a high incidence of anomalies of the central nervous system.

Foetal and Infant Loss and Morbidity.

Some aspects of this have already been discussed, but in the present section an attempt will be made to give an overall picture of the foetal and infant loss which occurs to the parents of diplegic patients.

It will be seen from Table 90 that of the 214 pregnancies to 75 women who gave birth to diplegic children, 9% ended in abortion. Of the 194 pregnancies which continued beyond the 28th week, one ended in stillbirth. 196 live-born /

TABLE 90

Foetal and infant mortality and morbidity amongst siblings of diplegic patients

	Number	Approx. %
Total number of pregnancies including those which resulted in the birth of the diplegic patients, to 75 women.	214	100
Aborted.	20	9
Total number of live and stillborn children (including twins).	197	100
Stillborn.	1	.5
Died in neonatal period.	6	3
Died in post neonatal period.	4	2
Survived first year of life.	186	94.5
Diplegic.	76	38
Mentally subnormal or disturbed (1 only had psychiatric symptoms).	13	7
Physically abnormal.	5	3
Total surviving apparently healthy offspring.	92	47
Total of offspring resulting from the 214 pregnancies, including twins and abortions.	217	100
Total of healthy survivors.	92	43

born infants (including twins) resulted from the other 193 pregnancies. Six of these, about 3%, died in the neonatal period and 4 more, about 2%, before the end of the first year. Of the 186 children who survived the first year (94.5% of the original 197 still and live born infants) 76 suffered from diplegia (38%), 15 were mentally subnormal or suffered from psychiatric disorders (8%) and 5 (approx. 3%) showed congenital physical defects, of some severity. Only 92 of the original 197 still and live born infants, or 47%, could be considered healthy, and it should be re-emphasised that this is almost certainly an underestimate of the number of mentally subnormal siblings of patients.

When the number of pregnancies is related to the number of healthy children, it will be seen that 43% only, of all pregnancies to the mother of a diplegic child, are likely to result in a healthy, mentally normal child. It is of some interest that of the 9 siblings considered to be feeble minded, 5 of them were born to parents who were themselves considered to be feeble minded, a finding of statistical significance. As has been noted, the association of abnormal siblings and abnormal parents in the same case, the association of abnormal siblings and neonatal and postneonatal deaths, and the association of abnormal parents and neonatal and postneonatal deaths occur with statistically significant frequency. On the other hand there is no significant association between the occurrence of abortions and that of abnormal parents, infant deaths or other abnormal siblings

in /

in the 75 cases studied.

If the abnormalities found in other siblings were frequently the result of birth injury, it might be expected that abnormal siblings would occur significantly more frequently in those cases in which there was a history of abnormal pregnancies and deliveries in addition to those resulting in the birth of the patient. In order to see if this were so, the 53 surviving siblings in families with a history of other abnormal pregnancies and deliveries were compared with the 57 born to mothers without a history of other abnormal pregnancies or deliveries (Table 91).

TABLE 91

Comparison of the Incidence of Abnormality in Siblings Born to Mothers with and without a History of Abnormal Pregnancy or Delivery

	No. of Mothers	No. of Sibs.	Number Abnormal	% Abnormal Siblings
With history of other abnormal pregnancies and deliveries.	27	53	12	23
With history of only other normal pregnancies and deliveries.	32	57	6	10

It will be seen that the proportion of abnormal siblings was more than twice as great in the cases in which mothers had other abnormal pregnancies, labours or deliveries as in those in which other pregnancies, labours and deliveries were normal, but in view of the small number of cases this difference is not great enough to be statistically significant. Moreover, when the proportions of abnormal siblings born after

normal /

normal and abnormal pregnancies, labours and deliveries are compared no significant difference is found.

Thus though the abnormalities present in the siblings of diplegic patients cannot be attributed to the effects of abnormal pregnancy, labour and delivery in a significant number of individual cases, there appears to be a tendency for mothers with abnormal other pregnancies to give birth to other abnormal siblings. Presumably other aetiological factors than birth injury must be present.

CHAPTER 7b

THE BIRTH HISTORIES OF DIPLEGIC PATIENTS

DESCRIPTION OF THE PREGNANCIES AND DELIVERIES OF THE 44 MATURE DIPLEGIC PATIENTS.

The 44 mature diplegic patients were classified (similarly to those born prematurely) into groups according to whether abnormalities of pregnancy and delivery were present or absent. Information was felt to be adequate and probably reliable in 41 of these. In three full information could not be obtained (Cases 114, 122 and 193). In Case 193 the child was known to have been a dissimilar twin with a birth weight of between 5 and 6 lbs., but neither in her case nor in the other two was the information adequate or reliable enough for tabulation.

No apparent abnormality of pregnancy or delivery.

Fifteen patients were classified as having no apparent abnormality or pregnancy or delivery. Six of the 15 mothers were over the age of 35 at the time of birth. Only 5 of the 15 children were first born. In all cases the health of the mothers was considered by the attendant obstetrician to be normal during pregnancy. In no cases was vomiting more than slight even in the early weeks. (Table 92)

Two of the patients were delivered at an estimated 39 weeks of gestation, and three at an estimated 41 weeks. The remainder were delivered within a few days of the expected date. The duration of labour varied from 3 hours in Case 205 to 25½ hours in Case 161. All the deliveries were spontaneous and by the vertex, usually under chloroform or nitrous oxide or pethidine analgesia. The condition of the patients /

patients was stated or implied to be good in 12 cases. In Case 12 the child was cyanosed for some hours after delivery. In Case 103 the child had a large caput and was cyanosed at birth. There was "slight apnoea" but thereafter the child seemed to be normal. The birth weights varied from 6 lbs. 8 ozs. in Cases 40, 133, 153 and 205 to 9 lbs. 8 ozs. in Case 96. All but the latter were in the average range.

Pregnancy abnormal but labour and delivery normal.

Eight patients were found to have abnormal pregnancies but normal labours and deliveries. All the children in this group were delivered spontaneously by the vertex after labours lasting between 2 hours in Case 10 and 23 hours 35 minutes in Case 22. In Case 46 the exact duration of labour was unknown but it was less than 24 hours. (Table 93)

The health of the mothers was said to be normal except in Case 10 in which the mother had had recurrent pyelitis prior to pregnancy and in Case 59 in which the mother, aged 42 years, was excessively obese and had had menopausal symptoms with scanty periods some months before conception. In Case 120 the mother had been noted by her doctor to be very weary during pregnancy, but no organic abnormality was found to account for this. Four of the mothers were 35 years old or more (Cases 10, 22, 59 and 120).

Four mothers had abnormalities in the first two trimesters of pregnancy, and in two of these (Cases 10 and 120) abnormalities were also present in the last trimester. Three mothers had abnormalities of the last trimester only (Cases

22 /

TABLE 92

Birth histories of diplegic patients - Mature

No apparent abnormality of pregnancy or delivery

Case No.	Social Class	Mat. age	No. of preg.	Maternal health.	Duration of labour hrs.	mins.	Delivery	Birth weight	Condition of child	Estimated gestation weeks	Placenta	Obstetric history (Type of delivery)
12	III	38	2	Good	4	0	Spont.vertex	7/¼	Fair	40	Normal	1 previous high forceps.
40	V	27	2	Good	10	0	Spont.vertex	6/8	Normal	40	Normal	1 previous and 1 subsequent normal.
47	III	40	4	Good	2	25	Spont.vertex	8/4	Normal	40	Normal	3 previous normal. 1 forceps. 1 miscarr.
62	II	39	1	Good	12	0	Spont.vertex	8/5	Normal	41	Normal	1 later prolonged labour.
76	I	31	4	Good	6	0	Spont.vertex	8/0	Normal	40	Normal	3 previous normal.
103	IV	26	1	Good	7	20	Spont.vertex	7/8¼	Cyanosed.	39	Normal	-
96	III	19	1	Good	4	45	Spont.vertex	9/8	Normal.	41	Normal	-
128	IV	36	4	V.anxious neurotic	6	0	Spont.vertex	8/0	Normal	40	Normal	3 previous normal.
133	III	27	2	Good	17	0	Spont.vertex	6/8	Normal	40	Normal	1 previous normal.
138	III	22	1	Good	Unknown		Spont.vertex	6/13	Normal	40	Unknown	1 subsequent delivery well.
153	IV	31	4	Toxoplasmosis.	9	0	Spont.vertex	6/8	Normal	40	Normal	3 previous well. 4 subsequent well.
161	V	24	1	Good apart from deafness.	25	30	Spont.vertex	7/5	Normal	41	Normal	2 normal. 1 died.
205	IV	36	8	Good	3	0	Spont.vertex	6/8	Normal	40	Unknown	1 forceps. 1 premature 5 previous normal.
206	II	30	2	Epileptic	7	0	Spont.vertex	7/4	Normal	40	Normal	1 previous and one subsequent normal.
207	III	44	2	Well but menopausal.	28	0	Spont.vertex	7/8	Normal	39	Normal	1 previous forceps.

(Details not available for Cases 114, 122 and 193)

22, 59 and 138). Case 10 passed a urinary stone during her pregnancy and had renal colic of some severity several times during her pregnancy in all three trimesters on other occasions. Case 46 attempted abortion with quinine and then by mechanical means (method not further specified) when 4 months pregnant. There was slight bleeding for some hours but pregnancy continued. Case 60 had a threatened abortion at three months and was confined to bed with moderate bleeding initially for about one week. Case 120 had very severe vomiting during the first three months of pregnancy. At six months she had a heavy fall downstairs with considerable bruising and intermittent pains afterwards which were taken to be "false labour pains" for some hours afterwards.

Four patients had pre-eclampsia. It was mild and in the last two months of pregnancy in Cases 59 and 138. It was moderately severe in Cases 22 and 118, and required early admission to hospital one week before term in the former. Cases 22 and 59 went into labour 4 and 7 weeks before the expected date and the birth weights were 5 lbs. 15 ozs. and 5 lbs. 13½ ozs. respectively. In Case 22 the child was limp at birth, showed some minutes' apnoea and was very flaccid. The placenta showed infarcts in Cases 22 and 59, but in Case 138 infarcts were not observed. In Case 118 the child seemed healthy.

With the exception of Case 22, all the infants breathed immediately after birth and seemed normal. In Case 10 the infant was large, weighing 9 lbs. 8 ozs.

Labour /

TABLE 93

The birth histories of diplegic patients - Mature

Pregnancy only abnormal

Case No.	Social Class	Maternal age.	No. of preg.	Health of mother and Abnormality of preg.	Duration of labour hrs.	mins.	Delivery	Birth weight	Cond. of child.	Estimated gestation wks.	Placenta	Obstetric history.
10	III	35	2	Nephrolithiasis with urinary infection.	2	5	Spontan. vertex	9/8	Good	40	Unknown	1 previous normal.
22	III	38	2	Moderately severe pre-eclampsia.	23	35	Spontan. vertex.	5/15	Lungs poor.	36	Infarcts + +	1 previous prolonged labour. Forceps.
46	II	30	1	Attempted abortion 4 months.	Unknown		Spontan. vertex.	6/12½	Good	40	Unknown	1 subsequent normal.
59	V	42	2	Obese menopausal mother with pre-eclampsia (mild)	17	15	Spontan. vertex.	5/13½	Good	33	Infarcts +++ Postpartum haemorrhage	1 previous normal.
60	III	23	1	Threatened abortion 3 months.	3	0	Spontan. vertex.	8/0	Good	40	Normal	-
118	III	19	2	Moderately severe pre-eclampsia.	11	40	Pituitary induction. Spon.vert.	7/14	Good but slow to feed.	40	Infarcts ++	1 previous abortion. 1 subsequent normal.
120	III	35	3	Much vomiting first 3 mths. Very heavy fall at 6 mths.	14	0	Spontan. vertex.	6/12	Good	44	Unknown	2 previous normal
138	III	23	1	Mild pre-eclampsia.	18	40	Spontan. vertex.	8/5½	Good	43	Normal	-

Labour or delivery abnormal but pregnancy normal. (Table 94)

Ten patients were found to have had normal pregnancies but to have been delivered instrumentally, or to have had abnormalities of labour. Seven of the nine were first born. Only two of the mothers were more than 35 years old (Cases 10 and 34). The physical health of the mothers was considered to be good except in Case 66, in which the mother was obese and had some vomiting (not considered to be significantly abnormal) in the early weeks of pregnancy, and in Case 10 where nephrolithiasis was present.

Four patients were delivered precipitously by the vertex, in a matter of moments at term (Cases 10, 34, 55 and 154). The condition of the child was poor after birth in Case 34, in which a medical induction of labour was necessary, the infant being apnoeic and cyanosed. In Case 154 the patient was merely noted to be in "fair" condition after birth. In Cases 55 and 10 the condition of the babies was good. In three cases the birth weight was between 7 and 7½ lbs. In Case 10 the female child weighed 9 lbs. 8 ozs.

Labour was prolonged on account of uterine inertia in three cases (Cases 56, 65 and 200). In Case 56 labour pains were intermittently present, and sometimes severe for 120 hours. In Case 65 pains were present for 72 hours and in Case 20^0 for 84 hours. Cases 56 and 200 were eventually delivered spontaneously by the vertex. In the former the condition of the child was good, but in the latter there was asphyxia pallida and a prolonged period of apnoea. The placenta /

placenta showed heavy infarction. Case 65 was delivered by high forceps in good condition and found to have a birth weight of 9 lbs. 8 ozs., though was of only 40 weeks' gestation. Case 66 and Case 104 also had uterine inertia during labour. The former was delivered by forceps on account of maternal and foetal distress after the head had been rotated from its persistently occipito posterior position. The child was in good condition. In Case 104 the onset of labour had been very gradual at an estimated 43 weeks' gestation and its duration is therefore difficult to estimate accurately. After more than 24 hours with poor irregular pains a mid-cavity forceps delivery was performed and an infant weighing 7 lbs. 4 ozs. delivered in quite good condition at an estimated 40 weeks' gestation.

In Case 18 high forceps delivery was performed after a labour of 3½ hours because of foetal distress. The condition of the child after birth was poor and apnoea lasted for about 10 minutes. The child weighed 5 lbs. 12 ozs. and had an estimated gestation of 37 weeks.

Pregnancy and labour or delivery abnormal.

Eleven patients had abnormalities in pregnancy and labour or delivery (Table 95). Two of the 11 mothers were over the age of 35 years. All the mothers were considered to be healthy from the obstetrical point of view, except Case 42 in which the mother, in poor social conditions, suffered from recurrent respiratory infections and was of very poor intelligence, and in Case 75 in which the mother had /

TABLE 94

The birth histories of diplegic patients - Mature

Labour or Delivery only abnormal

Case No.	Social Class	Mat. age	No. of preg.	Maternal health	Duration of labour hrs. mins.	Abnormality of labour.	Delivery	Birth weight	Cond. of child.	Estim. Gest. wks.	Placenta	Obstetric history
18	I	29	1	Good	3 30	Foetal distress	High forcep delivery.	5/12	Poor. Apnoeic	37	Normal	-
34	III	36	4	Good	minutes	Med. induction (ergot) precipitate.	Spontan. vertex	7/0	Poor. Did not cry.	40	Unknown	3 previous (1 forceps, 1 precip. labour, 1 stillbirth placenta previa.)
55	II	34	2	Good	0 15	Precipitate.	Spontan. vertex	7/7	Normal	40	Unknown	1 previous premature after prolonged labour, 1 subsequent high forceps delivery.
56	III	28	1	Good	120 approx.	Prem. rupture membranes. Uterine inertia.	Spontan. vertex	6/9	Good	43	Infarcts +++	1 subsequent normal.
65	IV	24	1	Good	72 0	Uterine inertia.	High forcep delivery.	9/8	Good	40	Normal	-
66	II	24	1	Vomiting and obese	12 0	Inertia. Mat. and foetal dis. Presented occip-ito posterior.	Rotation and forceps ex-traction.	6/8	Good	40	Unknown	1 subsequent forceps delivery.
104	III	28	1	Good	24+	Uterine inertia	Mid cavity forceps. delivery.	7/4	"Quite good"	40	Unknown	1 subsequent normal delivery.
154	IV	26	1	Neurotic	minutes	Precipitate.	Spontan. vertex.	7/0	Good	40	Unknown	1 subsequent normal.
200	V	17	1	Normal	82 0	Prolonged lab. Uterine interia	Spontan. vertex.	7/8	Asphyxia pallida	40	Unknown - post partum haemorrhage	4 subsequent normal.

had recurrent attacks of pneumonia, and judging by her halitosis (she refused examination) probably suffered from bronchiectasis.

In none of the cases in this group was the abnormality of pregnancy confined to the first two trimesters. Four mothers suffered from antepartum haemorrhage (Cases 25, 38, 42 and 126). In Case 25 the haemorrhage occurred at 36 weeks and was due to placenta praevia. The patient was delivered by Willett's forceps after artificial rupture of the membranes. Labour lasted two hours. The baby was limp and apnoeic at birth, weighed 5 lbs. 15 ozs. and was though to have had a gestation of approximately 36 weeks. The placenta showed very marked infarction. In Case 38 there was antepartum haemorrhage at 18, 28 and 39 weeks. On each occasion the haemorrhage was moderate in amount. External version was performed at the time of the last haemorrhage and shortly after the completion of this procedure labour commenced and continued intermittently for 48 hours. Much barbiturate and paraldehyde sedation was given but little analgesia. Delivery was spontaneous by the vertex. The child was very cyanosed and apnoeic after birth. In Case 42 the mother fell downstairs at an estimated 26 weeks' gestation, and when picked up was found to be bleeding vaginally. Bleeding continued for three days, and she refused offers of "abortion" at this time. The child was delivered by breech extraction after a labour of 24 hours with lack of progress since the child was presenting transversely. The child was apnoeic after birth and flaccid. The birth weight was 9 lbs. In Case 126 antepartum haemorrhage was /

had recurrent attacks of pneumonia, and judging by her halitosis (she refused examination) probably suffered from bronchiectasis.

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TABLE 96

The birth histories of diplegic patients - Mature

Pregnancy and labour or delivery abnormal

se o.	Social Class	No. of preg.	Mat. Age	Maternal health.	Abnormality of preg.	Abnormality of labour.	Duration of labour hrs.	mins.	Delivery	Bth. Wgt.	Cond. of child.	Estim. Gest.	Placenta	Obstetric history
5	V	1	19	Recurr. resp. infect.	Sev.antepart. haem. from placenta 36 wks.	Surgical induction.	2	0	Willet forceps	5/15	Limp and apnoeic	36	Infarcts ++	2 subsequent normal
8	IV	2	23	Good	Antepart.haem. at 18,28, and 39 weeks.	External version.	48	0	Spontan. vertex	7/4	Poor, apnoeic.	39	Unknown	1 previous and 2 subsequent normal.
2	V	5	28	Repeated resp. infect.	Fall at 6½ mths. followed by antepart. haem.	Malpresentation. Transverse lie.	24	0	Breech extraction	9/0	Poor, apnoeic.	41/42	Unknown	2 previous forceps. 2 previous normal.
7	I	1	37	Good	Threat. abort. 3 mths. Mod. sev. pre-eclamp. at 7 mths.	Inertia	16	0	Mid-cavity forceps.	6/4	Facial palsy. Otherwise well.	38	Normal	-
5	IV	3	32	Recurr. Pneum.	Pneum. & Pleur. during preg. 7/12.	Foetal distress.	17	20	Spontan. vertex	7/4	"Satisfactory".	40	Normal	2 previous normal.
1	II	1	25	Normal.	External vers. 4 times.	Prolonged transverse arrest.	53	0	Manual rot. Diff. mid-cavity for.	6/2½	Normal	40	Normal	1 subsequent normal.
6	I	1	32	Good	Mod. sev. pre-eclampsia.Antepart. haem. at 34 wks.	Pituitary Precipitate	minutes		Spontan. vertex	6/8	"Quite good considering".	40	Unknown	-
1	V	1	23	Good	Mod.sev. pre-eclampsia. External version at 7 mths.	Prem.rupture membranes. Dry labour.	27	15	Low forceps.	6/15	Good	40	Infarcts +	-
1	V	1	36	Normal	Vomiting 1 mth. Mod.sev. pre-eclampsia.	Prolonged uterine inertia. Foetal distress.	103	30	Mid-cavity forceps.	5/14	Poor Apnoeic	37	Infarcts +++	2 subsequent normal.
7	I	1	31	Normal	Sev. pre-eclampsia and hydramnios. Hyperpiesis.	Artif. rupture membranes. Prolonged labour.	72	0	Mid-cavity forceps.	6/12	Good	40	Unknown.	1 subsequent well. -
8	V	1	24	Good	Mod. severe pre-eclampsia.	Medical induction Foetal	6	40	Spontan. vertex	6/9½	Well	40	Many infarcts.	-

was a complication of moderately severe pre-eclampsia. It occurred at 34 weeks and blood loss was relatively heavy for three days. Pituitrin injections were given to induce labour at 40 weeks' estimated gestation, and a spontaneous vertex delivery was achieved after a precipitate labour lasting between 5 and 10 minutes. The condition of the child at birth was stated to be "quite good considering". The birth weight was 6 lbs. 8 ozs.

In addition to Case 124, five patients were born after pregnancies complicated by pre-eclampsia. In all the condition was classified as "moderately severe" in degree. Case 57 had a threatened abortion at three months and developed moderately severe pre-eclampsia at 7 months which necessitated admission to hospital 10 days before delivery. Mid-cavity forceps delivery was performed at an estimated 38 weeks' gestation after 16 hours' labour. The child had a facial palsy from the time of birth but otherwise appeared to be normal. The birth weight was 6 lbs. 4 ozs. Case 131 attempted abortion at 3½ months by taking large doses of quinine. She suffered from moderately severe pre-eclampsia in the last two months of pregnancy. External version was performed at seven months. After premature rupture of the membranes labour pains were irregular and a low forceps delivery was performed after a labour of 27 hours 15 minutes. The child was in good condition after birth and weighed 6 lbs. 15 ozs. The placenta showed marked infarction. In Case 141 the mother vomited excessively for the first month

of /

of pregnancy and developed moderately severe pre-eclampsia in the 32nd week. Labour was very prolonged owing to uterine inertia and foetal distress became apparent. After 103 hours and 30 minutes a mid-cavity forceps was performed at an estimated 37 weeks' gestation and an apnoeic child in poor condition extracted who weighed 5 lbs. 14 ozs. The placenta was heavily infarcted. Case 147 had severe pre-eclampsia and hydramnios of gradual onset. Surgical induction of labour by artificial rupture of the membranes on account of the toxaemia was followed by prolonged labour lasting 72 hours, after which time a mid forceps delivery was performed. The child's birth weight was 6 lbs. 12 ozs., and his condition was good at birth. Case 198 had excessive vomiting in the first 10 weeks of pregnancy and moderately severe pre-eclampsia later. Medical induction was performed at an estimated 40 weeks' gestation and after a labour of 6 hours 40 minutes the patient was delivered spontaneously by the vertex. The birth weight was 6 lbs. 9½ ozs. There were many placental infarcts.

In Case 75 the mother, a neurotic woman of low intelligence, suffered from persistent vomiting, and at seven months' gestation suffered from lobar pneumonia with pleurisy for 3 weeks. The child was delivered spontaneously at term after a labour of 17 hours 20 minutes, during which he had shown foetal distress. The condition at birth was rather unhelpfully noted as being "satisfactory". The birth weight was 7 lbs. 4 ozs. In Case 81 the child was subjected four times /

times to external version between the 28th and the 40th week of pregnancy. Labour occurred at an estimated 40 weeks' gestation and was prolonged for 53 hours on account of transverse arrest. After manual rotation a difficult mid-cavity forceps delivery was performed. The child's weight was 6 lbs. 2½ ozs. and its condition was stated to be normal.

The post-natal state and neonatal course of 40 mature diplegic patients. (Table 97)

Of the 14 patients in whom no history of any abnormality of pregnancy or of labour or delivery was elicited, 11 were apparently normal immediately after delivery and only one of these, Case 47, had an abnormal neonatal course consisting of a very marked loss of weight in the first week of 20 ozs. Three patients were unhealthy immediately after delivery. In Case 12 there was post-natal apnoea which lasted for some minutes, and the child's condition was noted to be "cyanosed and poor" even after resuscitation. Great poverty of movement and drowsiness were present in the first two weeks after delivery. In Case 40 the child was slow to cry after birth and seemed rather drowsy. During the neonatal period there was great difficulty in getting her to suck or swallow. In Case 103 the child showed no apnoea but was cyanosed after delivery and respirations were irregular and shallow for some hours so that oxygen was given - with resultant improvement. On the tenth day of life, however, the child suddenly became cyanosed and restless, with a bulging fontanelle and was considered to have had a cerebral haemorrhage. Apart from

Case /

Case 103, none of the patients in this group appears to have become anoxic during the neonatal period.

Of the 8 patients in whom pregnancy was abnormal but labour and delivery normal, 5 were apparently healthy immediately after delivery (Cases 10, 46, 60, 118 and 138). In all of these but Case 10 the subsequent neonatal course appeared to be normal. In the latter the child had a convulsion at the age of three days with loss of consciousness and was cyanosed for 48 hours thereafter. A cerebral haemorrhage was diagnosed and oxygen given for 48 hours. In Cases 60 and 118 the infants were noted to be rather quiet and slow to feed but were not considered to be abnormal at the time. Three patients were abnormal immediately after birth. In Cases 22 and 59 there was apnoea and the patients were cyanosed and limp, requiring resuscitation. In Case 22 there was much bruising of the head and great difficulty was experienced in making the lethargic infant feed. In Case 59 the infant showed oedema and jaundice in the first week and had some twitching of the limbs during this time. In Case 120 there was no neonatal apnoea but there was delay in crying and the tone and colour were poor for some hours after birth. Thereafter respiration persisted shallow and irregular, and the babe was restless and noisy for the first three days.

Nine patients gave a history of normal pregnancy but apparently abnormal labour or delivery. In 5 the patients were normal immediately after birth, and two of them had normal neonatal periods (Cases 55 and 56). In two the

neonatal period was complicated. In Cases 104 and 154 there were sudden cyanotic attacks on the third day in each case which were diagnosed as cerebral haemorrhages and which necessitated treatment with oxygen. Five patients were severely apnoeic after delivery (Cases 18, 34, 65, 66 and 200). In Case 18 the child was in a state of asphyxia pallida after a traumatic delivery and required much resuscitation. He remained very feeble for the first week of life and the left arm was noted to be flaccid at this time. In Case 34 the infant failed to breathe for some minutes after delivery though immediately after birth he was pink in colour. Respiration was very feeble for two hours and the child showed irregular respiration for 24 hours and thereafter had difficulty in sucking and swallowing. In Cases 65 and 200 apnoea was succeeded by "feebleness" or restlessness", and in Case 65 the child was generally hypotonic and in addition showed a completely flaccid right arm. In Case 66 the state of severe apnoea was followed by no complications and the child appeared to be thriving when discharged from hospital on the tenth day.

Nine patients gave a history of having a normal pregnancy and labour or delivery. In 3 of them (Cases 57, 131 and 198) the children appeared to be well immediately after delivery (though there was a slight lower motor neurone facial palsy in Case 57), and the neonatal courses were uncomplicated. In Case 147 the child appeared to be well immediately after delivery but had a "cerebral haemorrhage" not confirmed by lumbar puncture on the 7th day, from which he had apparently recovered /

TABLE 98

Neonatal health of patients suffering from diplegia - Mature

	Normal immediately after birth.		Abnormal immediately after birth.		
	Normal in neonatal period.	Abnormal in neonatal period.	Normal in neonatal period.	Abnormal in neonatal period.	Total
Pregnancy and deliv. normal.	11	1	0	3	15
Pregnancy only abnormal.	4	1	0	3	8
Labour or delivery only abnormal.	2	3	1	3	9
Pregnancy and labour or deliv. abnormal.	3	1	1	5	10
Total	20	6	2	14	42

recovered completely within 48 hours. In 5 patients the condition after birth was considered to be abnormal. One of these, Case 81, suffered from brief apnoea immediately after delivery and was cyanosed and limp for some hours. He responded to oxygen, however, and thereafter the neonatal course was apparently normal. In the other four cases the neonatal courses were abnormal. Case 25 had brief post-natal apnoea and respiratory movements were poor for three days, during the first two of which oxygen was required. In Case 38 there was great difficulty in swallowing and feeding and the child was very drowsy for the first week after post-natal apnoea lasting 15 minutes. Feeding difficulty was also encountered in Case 42 who also suffered from severe apnoea and was very slow to cry. Respiration was feeble in Case 75 for the first weeks after delivery, though only brief apnoea had been present. In Case 141 there was only brief apnoea but the child remained cyanosed and limp for some hours and was noted to be irritable, restless, fretful and very difficult to feed throughout his stay in hospital.

THE BIRTH HISTORIES OF 35 PREMATURELY BORN DIPLEGIC PATIENTS.

Since 27 of these patients had been born in hospitals and three in nursing homes the deliveries were relatively well documented in the majority and full case histories were available.

Of the 35 patients five were twins, including one pair of identical twins. In five cases there was no history of any /

any abnormality of pregnancy or delivery, apart from the fact that the children were born prematurely. In 8 cases pregnancy was abnormal but delivery normal. In 4 delivery only was abnormal, and in 17 both pregnancy and the actual delivery were abnormal. The births will be described according to whether pregnancy or delivery or both were abnormal.

Patients with history of normal pregnancy and delivery.(Table99)

There were 5 patients in this category (Cases 45, 98, 99, 177 and 183). All were first born. In Cases 45 and 177 the mothers were healthy. In Case 98 the mother was feeble minded and living in poor social conditions. Apart from obesity she was physically healthy. In Case 183 the mother, aged 39, had had infrequent scanty periods prior to conception, and following delivery periods did not return. She was considered to be menopausal. In Case 99 the mother had suffered from pulmonary tuberculosis for some years prior to conception, and though this was thought to be quiescent at the time of pregnancy she was readmitted to a Sanatorium shortly after delivery.

The pregnancies appeared to be normal in all cases apart from the premature onset of labour. Spontaneous delivery by the vertex occurred in all, though the membranes were deliberately ruptures during labour in Case 45, and an episptomy was performed in Case 177. In none of the 5 cases did sedation or anaesthesia appear to have been excessively heavy.

Four of the children breathed immediately after delivery. In /

In Case 99 apnoea after birth was present for some minutes. The child was very cyanosed. His birth weight was only 2 lbs. 12 ozs. and he required prolonged incubation. In Case 183 the child became abnormally cold three days after delivery and had to be admitted to hospital at that time where incubation was necessary. In Cases 45, 98 and 177 the neonatal period was uneventful.

Pregnancy abnormal but labour and delivery normal.

In seven cases the patients were prematurely born spontaneously by the vertex after abnormal pregnancies but normal labours (Table 100).

Three of the mothers were healthy up to the time of their pregnancies, though one of the four (Case 31) was feeble minded and had a husband of lesser rather than greater intelligence. The other four mothers were unhealthy. In Case 13 the mother had had a previous ovariectomy at the time of appendicectomy and during her pregnancy was suffering from pulmonary tuberculosis which required sanatorium treatment a few months after delivery. In Case 140 also the mother had had a previous ovariectomy and in addition a few years later a unilateral mastectomy. In Case 155 the mother suffered from chronic pyelitis before and during her pregnancy. In Case 186 the mother suffered from thyrotoxicosis which remitted during pregnancy but was exacerbated very shortly afterwards.

The abnormalities of pregnancy consisted of antepartum haemorrhage in four patients. In Case 140 there were three haemorrhages /

Birth histories of diplegic patients - premature

No apparent abnormality of pregnancy, labour or delivery

Case No.	Social Class	Mat. Age	No. of preg.	Maternal health	Drugs	Duration of labour hrs.	mins.	Delivery	Bth. Wgt. lbs. oz.	Condition of child.	Estim. Gest.	Placenta	Obstetric history
45	V	21	1	Small but healthy.	Pethedine. Nitrous oxide.	11	30	Artif. rupture of membranes. Spont. vertex.	4/7	Satisfactory	39	1 lb. healthy	-
98	V	33	1	Feeble-minded.	None	13	-	Spont. vertex. episiotomy.	3/14	Good	33	1 lb. 4 oz. No infarcts.	2 sub. normal 1 prem. 1 abortion.
99	III	24	1	Tuberculous.	Chloroform.	16	5	Spont. vertex.	2/12	Very feeble.	28	No note of infarcts.	-
177	III	20	1	Healthy.	Chloroform.	20	30	Spont. vertex. episiotomy.	4/12	Fair. Incubated.	32	No note of infarcts.	Three subsequent normal.
183	IV	39	1	Menopausal.	None.	3	-	Spont. vertex.	"Normal premature"		32	Infarcts +++ 1 lb. 12 oz.	-

TABLE 101

Birth histories of diplegic patients - premature

Delivery abnormal only

Case No.	Social Class	Mat. Age	No. of Preg.	Dur. of labour hrs.	mins.	Maternal health	Abnormality of labour	Delivery	Drugs	Bth. Wgt. lbs.oz.	Condition of child.	Estim. Gest.	Placenta	Obstetric history
41	V.	34	12	17	40	Healthy. Twin Preg. Patient first.	Breech present. Assist. breech extraction.		Chlorof.	3/13½	No apnoea. Incubated.	36	9 oz. lateral insertion.	5 prev. misc. 1 pre twin preg.2 abnor.c 3 normal 1 subnorma
63	III	31	4	4	-	Pul.Tub. Twin Preg. Pat. 2nd.	Breech present. Assist. breech extraction.		Chlorof.	3/13	No apnoea. Fair cond. Incubated.	40	No note.	2 prev.premature. 2 prev. abortions.
77	V.	16	1		4	Normal.	Precipitate labour.	Spont. vertex.	None.	3/15	Cyanosed, feeble,no apnoea. Incubated.	31	Normal	2 normal preg. subsequently.
53	IV.	42	4	1	-	Obese. Bronchit-ic.	Rapid	Spont. vertex.	None.	2/8	Very poor. Brief apnoea.	28	Unknown	3 previous of which 2 premature.
114	V.	18	3	9	20	Poor intell- igence.	Heavily sedated nembutal so as to be comatose.	Spont. vertex.	Nembutal	4/5	Brief apnoea. Sleepy prem. Incubated.	40	Normal	1st.child prec.labo 36 wks. 2nd. child prec. labour birth.
158	V	23	1	11	23	Normal	Assist. breech w. pituitrin. occipto post.		Chlorof.	4/13	Normal prem. twins except for talipes equinus varus.	40	2 lbs.	-
159	V	23	1	11	48	Normal	"		"	5/5½				

haemorrhages between the twenty-first and twenty-third weeks, the last occurring immediately before the onset of labour. In Case 31 a persistently brown discharge which contained frank blood from time to time was present from the fifth month of gestation following a vaginal haemorrhage until the onset of labour. In Case 13 the tuberculous mother was admitted to hospital with a diagnosis of bronchitis in premature labour immediately after she had had a very severe vaginal haemorrhage. In Case 155 the mother was suffering from pyelitis and mild pre-eclampsia at the time of the occurrence of her haemorrhage which occurred approximately 24 hours before delivery.

In Case 189 there was a history of attempted abortion at approximately four months' gestation. This resulted in vaginal haemorrhage but thereafter pregnancy was apparently normal until the onset of premature labour at 28 weeks with rupture of the membranes.

The thyrotoxic mother in Case 186 suffered from very severe pre-eclampsia necessitating admission to hospital for rest on two occasions prior to delivery. At the time of delivery the placenta showed extensive infarction. In Case 203 there was a history of the mother suffering from severe influenza with chest complications during the first two months of her pregnancy, which was apparently normal thereafter.

Pregnancy normal but labour or delivery abnormal. (Table 101)

Seven patients came into this category. Four were twins /

twins including one pair of similar diplegic twins (Cases 158 and 159). In Cases 41 and 63 the dissimilar twins were healthy.

In Case 114 the mother was feeble minded but otherwise apparently healthy. In Case 63 the mother had suffered from pulmonary tuberculosis for some years before she became pregnant, but it was thought to be quiescent at the time of pregnancy. In Case 53 the mother was excessively obese, bronchitic and menopausal when she conceived the child at the age of 42 years. She was a full-time cleaner of stairs during her pregnancy. In Case 77 the mother was only 16 at the time of conception and worked as a shop assistant until shortly before the time of delivery.

Labour was precipitate in two cases (Cases 53 and 77). In the latter its duration was only "a few minutes". In the former it lasted approximately one hour. The patients showed only brief apnoea in Case 53 and no apnoea in Case 77, though the condition of both children was considered to be very poor, and the gestations 28 and 31 weeks respectively.

Three of the twins (Cases 41, 63 and 158) were delivered by breech extraction after labours of between four hours (Case 63) and 17 hours (Case 41). The condition of all three was considered to be fair following delivery except that Case 158 showed congenital talipes equino varus deformities. In Case 159, the second born of the identical diplegic twins, delivery occurred spontaneously by the vertex, the position being occipito posterior.

In /

TABLE 100

Birth histories of diplegic patients - premature

Abnormality of pregnancy only

Case No.	Social Class	Mat. Age	No. of Preg.	Maternal health	Abnormality of pregnancy	Dur. of labour hrs.mins.	Drugs	Delivery	Bth. Wgt. lbs.oz.	Condition of child.	Estim. gest.	Placenta	Obstetric history
13	V	22	2	Prev.ovariotomy Pul.tuberculosis	Antepartum haem.29 wks.	5 45	None	Spont. vertex with episiotomy	3/8	Some mins. apnoea.Fair. Responded to nikethamide. Incubated for 6 wks.	29	15 oz. normal Lateral insertion	1 previous boy born spont. at term.
31	IV	18	1	Poor intelligence.	Dark disch. present until deliv.following haem 5th. month.	5 -	None	Spont. vertex	2/6	Some mins. apnoea.Fair. Incubated.	28	Unknown	1 sub. miscarriage. APH on sub. preg.
140	III	24	3	Prev.ovariotomy & mastectomy.	Much vomiting for 5 mths. Antepartum Haem. 21,31 & 33 wks.	21 30	Pethedine +++ Soneryl	Spont. vertex.	3/10	Poor.Prolonged apnoea. Very cyanosed.	33	Infarcted	2 previous normal children.
155	III	30	4	Recurrent pyelitis.	Pyelitis 2 mths. prior to deliv. Mild pre-eclampsia. Antepartum haem. day before deliv.	6 35	Gas & Air	Spont. vertex	5/8	Poor. Apnoea. Incubated.	40	1 lb. No infarcts noted.	Three previous. One precipitate and two normal deliveries.
186	II	35	2	Thyrotoxic.	Severe pre-eclampsia.	2 45	"Sedation"	Spont. vertex.	5/2½	Normal prem. Incubated for 48 hrs.	35	1 lb. 12 oz. Infarcts +++	First preg. complicated by pre-eclampsia. Surgical induction.
189	V	27	3	Healthy.	Attempted abort. 4 mths. Prem. rupture memb. at 28 wks.	18 40	Much trilene.	Spont. vertex.	3/7½	Apnoea, cyanosed. Incubated.	28	12 oz. No note	1 previous abortion. 1 previous normal delivery.
203	II	22	1	Healthy	Severe influenza in first 2 mths of preg. with pulmonary complications.	5 30	Gas & Air	Spont. vertex.	3/-	Normal prem. Prolonged incubation.	29	Unknown	None.

In Case 114 the mother was excessively sedated with "Nembutal" so that she became alternately comatose and delirious and was quite unmanageable. The child was delivered spontaneously by the vertex after a four-hour labour and was apnoeic for a few minutes after delivery. Thereafter except for excessive drowsiness no abnormality was noted.

Pregnancy and labour or delivery abnormal. (Table 102)

Fifteen mothers had both abnormal pregnancies and labours or deliveries. Thirteen were considered to be in normal health prior to pregnancy, though in Case 156 the mother was of very poor intelligence.

In Case 32 the mother had suffered from recurrent attacks of pyelitis before she became pregnant and the condition was present during pregnancy. The mother in Case 157 had a large goitre and was mildly thyrotoxic. In addition she had severe anaemia which was resistant to iron therapy.

Five mothers had threatened or attempted abortion during pregnancy. In Cases 61 and 181 attempted abortions in the early months were followed by antepartum haemorrhages immediately prior to the onset of labour at an estimated 30 and 32 weeks respectively. In Case 181 the child was delivered spontaneously by the vertex after a labour of 8 hours 50 minutes together with a foetus papyraceous. In Case 61 the cord prolapsed during labour and the child was extracted by the breech. Both infants were apnoeic and required resuscitation and prolonged oxygen therapy. In Cases 57, 172 and 174 abortion threatened during the first three /

three months. In Case 57 there was mild toxaemia from the seventh month of pregnancy until term at which time mid-cavity forceps delivery was required to deliver the child owing to uterine inertia. The birth weight was 5 lbs. 2 ozs., and severe neonatal apnoea was present. In Case 172 pregnancy was uneventful after two episodes of bleeding at the first and second months of pregnancy, but labour was very prolonged on account of contracted pelvis and the very small child was in poor condition after birth though he breathed at once. In Case 174 vaginal haemorrhages occurred at the 10th and 29th weeks of pregnancy from placenta praevia and internal version and assisted breech extraction were performed as it was believed that the chances of the child were very small in any case, after a labour of 24 hours during which time haemorrhage had continued. The child was severely apnoeic and was in very poor condition.

Including the four cases (Cases 57, 61, 174 and 181) described above who had vaginal haemorrhage within the first three months and again in the last three months of pregnancy, nine patients had antepartum haemorrhages and these were the commonest abnormalities of pregnancy in this group. The five cases in whom antepartum haemorrhages occurred only in the last three months were Cases 2, 3, 83, 152 and 157. In Case 157 these occurred at the 27th and the 32nd weeks and complicated pre-eclampsia. At the time of the second haemorrhage much morphia was given and then a Caesarean section was performed to deliver an infant in poor condition and /

TABLE 102

Birth histories of diplegic patients - premature

Pregnancy and delivery abnormal

Case No.	Social Class	Mat. Age	No. of Preg.	Maternal health.	Abnormality of pregnancy.	Dur. of labour hrs.mins.	Abnormality of labour.	Drugs	Delivery	Bth. Wgt.	Condition of child	Estim. gest.	Placenta	Obstetric history
2	III	31	3	Good.	Much vomiting. Haem. 3 days before deliv.	84 -	Prolonged.	Trilene	Spont. vertex.	3/3	No apnoea. Cyanosed. Incubated	29	1 lb. ⅔ very pale.	1 prev. misc. 1 prev. normal deliv. 2 wks.
3	II	34	2	Good.	Haem. 8 mths. Morphia and med. induction.	- 15	Precip.followed forceps deliv. contracted outlet	Chloro.	Low forceps	4/15	No apnoea Well.	36	Unknown	1 prev. normal delivery.
32	IV	27	2	Recurr. Pyelitis	Pyelitis dur. pregnancy.	10 30	Breech pres. given ½ gr. morphia.	Chloro.	Spont. breech deliv.	3/-	5-10 mins. apnoea. Shocked & incubated.	30	Unknown	1 prev. normal deliv. 1 prev. miscarriage.
57	I	37	1	Good.	Threatened.abort. 3 mths. Mild tox. from 7th.to 9 mths.	16 -	2nd. stage delay.Uterine inertia.	Omnipon Hyoscine Chloro.	Mid-cav. forceps.	5/2	Prolonged apnoea.Cyan. and feeble.	40	Unknown	-
51	III	23	1	Healthy.	Much vomit. Persis. fainting. Premature rupture membranes.	- 30	Precipitate delivery	Gas & air. Hyoscine	Spont. vertex.	4/-	Fair. No apnoea. Incubated.	32	No note	1 subsequent normal delivery.
61	IV	32	2	Healthy.	Much vomit. 2 mths. Beaten up 2 mths. preg.Attemp. abor. 3rd. & 4th.mths. Antepart. haem 6 mths. foll. assault	3 40	Prolapsed cord. Breech presentation,	Local	Assisted breech.	3/5	Brief apnoea 1-2 mths. Incubated.	30	1 lb.	1 previous high forceps delivery.
83	II	33	1	Normal.	Haem. and pains at 28 weeks.	47 -	Foetal heart disappeared.	Pituitrin.	Fundal pressure morphia pituitrin	2/14	Fair. No apnoea. Cyanosed. Incubated.	29	1 lb. retro-placental clot.	1 normal subsequent delivery.
124	III	37	2	Normal.	PET.	72 -	Prolonged labour.	Unknown	Spont.	3/8	Fair, no apnoea. Incubated.	32	Unknown	1 previous normal.
152	II	34	2	Healthy.	Haemorrhage when 7 mths. preg.	3 45	Artif. rup. membranes. Pethedine Breech pres.	Pethedine + Chloro.	Assisted breech.	3/11	Poor. apnoea. Incubated.	32	Old retro-placental clot.	1 previous premature deliv. 2 abortions.
156	V	21	1	Low I.Q.	Severe vomiting 3 mths. Severe pre-eclampsia.	37 25	Bougie induct. Inertia.	Luminal	Spont. deliv.	4/8½	Normal prem. Slight cyanosis	37	1 lb. many infarcts.	One subsequent normal delivery.
157	IV	34	3	Goitre. Anaemia.	Pre-eclampsia. Haem 27 & 32 wks. Morphia ++	-	-	Ether	Caesar. Section	5/-	Poor. Brief apnoea. Incubated.	32	No note	Four normal deliveries previously.
163	III	33	3	Healthy.	False labour,pains 1 wk. before deliv. ext.version 24 hrs. before delivery	mins.	Precipitate	None	Spont. vertex	4/-	Normal premature.	36	No note	Two previous normal deliveries.
172	III	32	1	Normal	Bleeding 1st. & 2nd. mths. of preg. APH prior to deliv.	72 -	Contracted pelvis. Prolonged labour.	None	Spont. vertex	2/12	Poor but breathed at once.	32	No note	2 previous abortions.
174	IV	23	3	Normal	Bleeding 2½ & 7 mth placenta previa.	24 -	Internal version. Leg brought down.	Chloro.	Assist. breech.	3/-	V.poor marked apnoea s.mths.	29	No note	2 pr.with APH, 3 sub., 2 APH,1 precip. deliv.
181	III	30	3	Normal	Hyperemesis 3 mths. attemp.abor.3½ mths (quinine 50 gr) Some bleeding.Twin preg. Antepart.haem.3 days before delivery.	8 50	Spont. with foetus papyraceous. (patient first)	None	Spont.	2/8½	V.poor Apnoea 7 mins.	32	Infarcts++	1 prev.normal deliv. 1 prev. abortion.

and weighing 5 lbs.

In Case 83 haemorrhage occurred immediately before the onset of labour at an estimated 28 weeks. The foetal heart disappeared after the mother had been in labour for 45 hours, and since the child was believed to be dead much morphia was given and the child expelled by fundal pressure. Though very small he was in fair condition and breathed at once. Antepartum haemorrhage occurred at 28 weeks also in Case 152. After artificial rupture of the membranes a labour of $3\frac{3}{4}$ hours ensued during which much pethidine was given. Breech extraction was performed and an apnoeic, very premature infant weighing 3 lbs. 11 ozs. was delivered.

Of the 9 cases in which antepartum haemorrhage occurred the infant was apnoeic in 6 after delivery. Apnoea was severe in Cases 57, 157, 174 and 181, and less severe in Cases 152 and 61. The condition of all the apnoeic infants was considered to be poor and in two of the remaining three (Cases 2 and 3) it was considered to be fair, though prolonged incubation and oxygen was required in the former. In Case 83 the child's condition was thought to be poor in spite of his breathing at once after birth, and prolonged incubation with oxygen was required.

In Case 32 the mother suffered from a recurrence of her pyelitis during pregnancy and premature labour began at 30 weeks. During her labour of $10\frac{1}{2}$ hours morphia was given as initially her pains were attributed to the pyelitis. A spontaneous breech delivery occurred of a very small severely apnoeic /

apnoeic child, weighing 3 lbs., who required prolonged incubation and oxygen.

In Case 51 the mother had extremely severe hyperemesis during the first three months of her pregnancy, and from the first two weeks of gestation suffered from as many as 15 or 20 fainting attacks each day. The child was delivered spontaneously by the vertex after a precipitate labour at an estimated gestation (which the mother could date accurately from coitus) of 32 weeks. She breathed at once but respiration was very shallow and incubation and oxygen were required for one week.

Apart from Case 157 in which pre-eclampsia was complicated by antepartum haemorrhage, two other mothers suffered from toxaemia. In Case 124 the mother aged 37 had moderately severe pre-eclampsia and went into labour at the 32nd week of pregnancy. There was severe uterine inertia for which repeated injections of morphia and pituitrin were given, and after 72 hours spontaneous vertex delivery of a child weighing 3 lbs. 8 ozs. in fair condition occurred. In Case 156 the unintelligent mother had severe vomiting in the first three months of her pregnancy and developed severe pre-eclampsia in the last three months. An aritificial bougie induction of labour was performed at 37 weeks, but uterine inertia ensued and delivery occurred only after 37 hours in labour. The child was slightly cyanosed after delivery but was not apnoeic and weighed 4 lbs. 8½ ozs.

Details of the condition of the placenta were available in /

in only one of the three cases with pre-eclampsia, and in that (Case 156) severe infarction was present.

The post-natal state and neonatal course of 35 premature diplegic patients.

In the large Table 103 are shown the available clinical details of the 35 prematurely born patients during the first months of life. They are grouped according to whether pregnancy and labour or delivery were normal or abnormal.

It will be seen that three of the 5 patients in whom both pregnancy and delivery were uncomplicated were apparently normal immediately after birth and during the neonatal period. One (Case 183) was hypotonic and lethargic during the neonatal period and required incubation for some weeks, which was hardly surprising in view of the fact that his birth weight was only 3 lbs. 8 ozs. and he was allowed to become chilled after delivery. Only one patient in this group (Case 99) was abnormal immediately after birth. He suffered some minutes' apnoea and required incubation and oxygen for some weeks during which he was noted to be hypotonic and lethargic.

Of the 7 patients in whom pregnancy was abnormal though the labour and delivery were uncomplicated, two were apparently normal immediately after delivery, and five were unhealthy. In both those who appeared to be healthy immediately after birth there were complications in the neonatal period; Case 186 was allowed to become cold after delivery and required incubation for 48 hours though his birth weight was 5 lbs. $2\frac{3}{4}$ ozs. He showed marked poverty of movement, hypotonia and difficulty in sucking and swallowing throughout the neonatal period. /

period. In Case 203 the child weighed only 3 lbs. at birth but breathed at once. Respiration was shallow, however, and the child very feeble. Incubation and oxygen were required for three weeks, and retrolental fibroplasia became apparent some months later.

One child in this group (Case 155) weighed exactly 5 lbs. 8 ozs. at birth and was severely apnoeic after delivery, requiring strenuous efforts to resuscitate him. He was given oxygen for 24 hours and then incubated, but responded well and his neonatal course was quite normal thereafter.

Four patients were abnormal immediately after birth and abnormal in the neonatal period afterwards (Cases 13, 31, 140 and 189). In Cases 140 and 189 there was severe post-natal apnoea and in Cases 13 and 31 less severe neonatal apnoea. All four required prolonged incubation, and all except Case 140 prolonged oxygenation on account of persistently poor respiratory movement and the occurrence of cyanotic attacks. In Cases 13 and 189 retrolental fibroplasia developed later in association with severe diplegia.

In Case 157 the child's birth weight was 5 lbs., but there was apnoea after birth and he was noted to be very feeble and cyanosed. After initial resuscitation oxygen was not required but he was incubated for three days and ran a sub-normal temperature for three weeks. Poverty of movement was noted. Cases 174 and 181 (the latter a twin) weighed 3 lbs. and 2½ lbs respectively at birth. They were both apnoeic immediately after delivery and required resuscitation followed /

TABLE 103

The Birth Histories of Diplegic Patients - Premature

The Neonatal Period

Case No.	Bth. Wgt.	Estim. Gest.	Apnoea	Condition	Resuscitation	Incubation	Oxygen	Complications
45	4/7	39	No	Lively	No	No	No	None
98	3/14	33	No	Good for small size.	No	No	No	None
99	2/12	28	Some min.	Feeble. Did not cry for 3½ days.	No	8 weeks	5 weeks.	Floppy and lethargic. Very poor respiratory movements for 10 days. Tube fed.
177	4/12	32	No	Cried well.	No	No	No	None.
183	3/8	32	No	Cried well.	No	Not for 2 days then became very cold and incubated for 4 weeks.	No	Very still floppy baby. Difficulty in feeding.
13	3/6½ (3rd.day)	29	Some min.	Feeble. Poor chest expan.	O_2 Nikethamide	For 6 weeks.	5 weeks.	Persistent atelectasis. Resp. infections. Severe anaemia requiring transfusions.
31	2/6	28+	Some min.	Shocked. Feeble.	O_2 Nikethamide Lobeline	For 6½ weeks.	3 weeks.	Severe cyanotic attack aged 6 days. Very difficult to feed. Floppy.
140	3/10½	33	10 min.	Flaccid. Cyanosed.	O_2 Vit K. Nikethamide	For 2 weeks.	3 days.	None after initial resuscitation. Noted to have bilateral talipes equinus varus.
155	5/8	40	Prolonged. Severe.	Cyanosed and grey colour. Very feeble.	O_2 Nikethamide	24 hours.	1 day	None. Good recovery.
186	5/2¾	35	No	Good but became cold after delivery.	No	48 hours.	No	Very still baby. Difficult to feed.
189	3/7½	28	Severe	Cyanosed and feeble.	O_2 Nikethamide	3 weeks.	2 weeks.	Mild cyanotic attacks in 1st.week. Inguinal hernia. Retrolental fibroplasia.
203	3/0	29	No	Quite good cond. for sick premature child.	No	3 weeks.	3 weeks.	Slow feeding. Poverty of movement. Floppy. Retrolental fibroplasia.
41 (twin)	3/13½	36	No	Fairly good.	No	No	No	Diarrhoea and anaemia.
53	2/8	28	Yes Brief	Very feeble. Irreg. Resp.	Slapping	No	No	None though difficult to feed. Kept at home.
63 (twin)	3/13	40	No	Shallow irregular resp. but good colour and tone.	No	Yes. Admitted to hosp. aged 5 days.	2 days.	None.
77	3/15	31	No	Good	No	1 week	4 days	Cyanotic attacks on 2nd. day. Feeding difficulty.
114	4/5	?40	Yes Brief	"Rather poor"	No	5 days	5 days	Slight vomiting.
158 twins	4/13	40	No	"Quite good"	No	No	No	None
159	5/5½	40	No	"Quite good"	No	No	No	None
2	3/3	29	No	Lively but very premature.	No	2 weeks	5 weeks	2 cyanotic attacks in 1st. week. Retrolental fibroplasia. Poor chest expan. Resp. Infecti
3	4/13	36	No	Cyanosed but not ill.	No	No	No	None except he was a very still baby.
32	3/0	30	5-10 mins.	Cyanosed and flaccid.	Coramine into cord. Oxygen.	For 12 weeks.	For 10 weeks.	Jaundice. Valgus deformity of left foot. Very feeble. Pus from umbilicus. Retrolental fibroplasia.
51	4/0	32	No	Fair. Shallow reg. respirations.	No	1 week	1 week	Persistent atelectasis (pneumonia) "Cerebral attacks".
57	5/2	40	Prolonged.	Cyanosed and feeble.	O_2	2-3 days	2-3 days	Feeble for a day or two. Thereafter well.
61	3/5	30	1-2 mins.	Cyanosed and limp.	O_2	Few days	2 days	No.
83	2/14	29	No	Cyanosed and limp.	O_2 Nikethamide	3 weeks	11 days	Very feeble. Poor respirations.
124	3/8	32	No	Cyanosed and flaccid.	O_2	About 4 weeks.	About 8 days.	Slow weight gain. Very difficult to feed.
152	3/11	32	Frief	Cyanosed and very flaccid.	O_2 Endotracheal Lobeline into cord.	2 weeks	3 days	None
156	4/8½	37	No	Slightly cyanosed.	No	No	No	Did not gain any weight for 3 weeks. Very feeble.
157	5/0	30	Some mins.	Cyanosed.	O_2. Injections of coramine.	3 days	No	Subnormal temperature for 3 weeks and rather feeble and quiet.
163	4/0	36	No	Good	No	No	No	Very sleepy. Poor feeder.
172	2/12	32	No	Poor. Limp. Feeble resp.	No	No	No	Gastro-enteritis aged 3 weeks.
174	3/0	28-29	Some mins.	Feeble. Cyanosed.	Put under cold tap.	Between one and 3 weeks.	For some days.	Quiet still baby. Very difficult to feed without choking.
181 (twin)	2/8½	32	7 mins.	Feeble. Cyanosed.	Lobeline. Coramine. oxygen.	7 weeks	6 weeks	Very quiet. Cyanotic attacks 5 times in first week. Persistent atelectasis. Pneumonia.

followed by incubation and oxygen. They both had cyanotic attacks in the early weeks, showed poverty of movement, and Case 181 developed neonatal pneumonia. The other four cases (Cases 83, 124, 156 and 172) were all cyanosed after delivery and were limp, but none of them was apnoeic. Cases 83 and 124 had persistently poor chest expansion and required prolonged incubation and oxygen. Case 156, whose birth weight was 4 lbs. 8½ ozs., was feeble and failed gain weight for three weeks. Case 172 was thriving well until he developed gastro-enteritis at the age of three weeks.

Of the 7 patients in whom labour or delivery was complicated but pregnancy was normal, 5 were apparently healthy immediately after birth, and three of these, all twins (Cases 41, 158 and 159) had normal neonatal periods. In Cases 63 (a twin) and 77 the children appeared to be in good condition immediately after birth, but the former became cold after five days and had to be admitted to hospital for incubation, and the latter, whose birth weight was 3 lbs. 15 ozs., suffered from cyanotic attacks from the second day onwards and had great feeding difficulty. Two patients were in poor condition immediately after delivery. In Case 53 the infant was born with a birth weight of 2 lbs. 3 ozs. at home. Though he suffered from brief apnoea after delivery and shallow irregular respiration for some weeks afterwards, the neonatal period was normal apart from some minor feeding difficulty though he was nursed at home in poor social conditions by a mother of no more than low average intelligence.

In /

In Case 114 there was apnoea of short duration immediately after delivery, and the child's condition was described as "rather poor" though the birth weight was 4 lbs. 5 ozs. Respiration was very shallow for five days during which time incubation and oxygen were given, but after this time the further progress was normal.

Of the 15 premature infants in whom pregnancy and delivery had been abnormal, only 5 appeared to be healthy at birth and only three of these had normal neonatal periods (Cases 3, 156 and 163). All these patients had birth weights of over 4 lbs. Cases 2 and 51, with birth weights of 3 lbs. 3 ozs. and 4 lbs. respectively, both appeared to be normal immediately after birth but suffered from complications during the neonatal period. The former persistently had very poor chest expansion and required incubation for seven weeks and oxygen for five. He had two severe cyanotic attacks in the first week of life. Later on recurrent chest infections occurred and retrolental fibroplasia was observed before he was 10 weeks old. In Case 51 there was persistent pulmonary atelectasis, and "cerebral attacks" occurred during the first four days of life. For the first weeks after delivery the child was nursed in oxygen and incubated.

Ten patients appeared to be in poor condition immediately after delivery in this group. In all the neonatal course was complicated. Case 32 was persistently jaundiced during the first week of the neonatal period, and noted to be very feeble. She was severely apnoeic after delivery and required oxygen

and /

and incubation for ten weeks. In Case 57 post-natal apnoea was severe and respiration poor for three days, but thereafter the neonatal course was normal. In Case 152 there was brief apnoea and after resuscitation oxygen and incubation were required, but apart from persistently poor chest expansion no other complications were encountered.

Following uncomplicated labours and deliveries the patients were considered to be in quite good condition in two cases (Cases 186 and 203), neither of whom was apnoeic. In the other five cases their condition was poor and there was apnoea, which was severe in three, and lasted "for some minutes" in the other two. All the children required incubation, and all but one of them (Case 186) whose birth weight was 5 lbs. 2¾ ozs. needed oxygen.

CHAPTER 7c

CONSIDERATION OF THE SIGNIFICANCE OF ABNORMALITIES OF PREGNANCY, LABOUR AND DELIVERY IN PATIENTS WITH CONGENITAL DIPLEGIA

Early Pregnancy.

The abnormalities of the first two trimesters of pregnancy are shown in Tables 104a and 104b . The most common abnormalities of the first six months of pregnancy in both the mature and premature groups was incomplete abortion, either accidental or induced. It will be observed, however, that the proportion of prematurely born patients with a history of incomplete abortion, 7 in 34 cases (approximately 21%) of whom full details were known, was considerably higher than the proportion of mature patients with a similar history, 4 in 40 (~~approximately~~ 10%). Amongst the mature patients Cases 46 and 60 are of some interest, and amongst the prematurely born patients Cases 31 and 189, as in them attempted or threatened abortion was the only apparent abnormality of pregnancy, labour or delivery, apart from premature rupture of the membranes in the latter two. The condition of both the premature children was poor, however, though that of the mature children was good.

In the remaining two mature patients (Cases 38 and 131) and the other premature patients (Cases 61, 57, 140, 172, 174 and 181) there were other abnormalities of pregnancy, labour or delivery. In Case 38 of the mature group and in Cases 61, 140, 172, 174 and 181 of the premature group there were recurrent haemorrhages in later pregnancy and in Case 131 of the mature and in Case 57 of the premature group there was later /

Birth birth histories of diplegic patients

Disorders of the first two trimesters

	Case No.	Mat. Age	No. of Preg.	Maternal health	Pregnancy	Labour and Complications	Dur. of labour hrs.mins.	Delivery	Drugs	Condition of child	Birth Wgt. lbs.	oz.
A. MATURE	38	23	2	Healthy.	Haemorrhage at 18,28,39 weeks.	Ext.version at onset of labour. Prolonged.	48 -	Spont.vertex	None	Poor. Apnoea	7	4
	46	30	1	Good.	Attempted abortion at 4 months.	Normal	Unknown	Spont.vertex	None	Good	6	12½
	60	23	1	Good.	Threatened abortion at 3 months.	Normal	3 -	Spont.vertex	None	Good	8	-
	131	23	1	Good.	Attempted abortion 3½ mths. Pre-eclampsia. External version at 9 months.	Dry labour.	27 15	Low forceps	Chloro.	Good	6	15
B. PREMATURE	31	18	1	Poor intelligence.	Haemorrhage at 5 mths. Premature rupture of membranes.	Normal premature.	5 -	Spont.vertex	None	Fair	2	6
	57	37	1	Healthy.	Threatened abortion at 3 mths. Mild pre-eclampsia 7-9 mths.	Uterine inertia.	16 -	Mid cavity forceps	Chloro.	Prolonged apnoea.	5	2
	61	32	2	Healthy.	Assaulted at 2 mths. Attempted abortion 3rd. & 4th. mths. Antepartum haem. at 29 weeks.	Prolapsed cord.	3 40	Assisted breech	None	Apnoea	3	15
	140	24	3	Previous ovariectomy.	Hyperemesis. Haem. at 21,31 and 33 weeks.	Normal	21 30	Spont.vertex	Pethedine Soneryl	Severe apnoea.	3	10
	172	32	1	Healthy.	Threatened abortion at 1 & 2 mths. Antepartum haem. placenta previa.	Prolonged labour.	72 -	Spont.vertex	None	Poor	2	12
	174	23	3	Healthy.	Bleeding at 2½ & 7 mths. Placenta previa.	Internal version.	24 -	Assisted breech	Chloro.	Severe apnoea.	3	-
	181	30	3	Healthy.	Twin preg. Hyperemesis. Attempted abortion 3½ mths. Antepartum haem.	Normal twin.	8 50	Spont.vertex with foetus papyraceous.	Chloro.	Very poor. Severe apnoea.	2	8½
	189	27	3	Healthy.	Attempted abortion at 4 mths. Prem. rupture membranes at 28 weeks.	Normal premature.	18 40	Spont.vertex	Trilene	Apnoea.	3	7½
	203	22	1	Healthy.	Severe influenza with secondary chest complications first 2 mths. pregnancy.	Normal premature.	5 30	Spont.vertex	Gas & Air	Fair	3	-

later toxaemia.

In Case 203 of the premature group severe influenza with pulmonary complications occurred in the first two months of pregnancy, but this was the only abnormality of pregnancy, labour or delivery in this case, though the condition of the child at delivery was considered to be only "fair".

From the above it is apparent that while eight (approximately 24%) of the 34 patients in the premature group in whom full details were known had apparently abnormalities of pregnancy in the first six months, in only two cases were these the only apparent causes of possible damage to the foetus prior to delivery. In the mature group only two patients gave a history of pregnancy and delivery which was abnormal only in the first ~~six~~ six months of gestation, and a further two of abnormalities in the first six months of pregnancy and in later pregnancy and delivery.

Thus apparent abnormalities of early pregnancy, most commonly incomplete abortion, are present in approximately 16% of all diplegic patients, including approximately one quarter of premature patients. On the other hand abnormalities of early pregnancy are relatively seldom unaccompanied by disorders of later pregnancy, labour and delivery. It is theoretically only possible for apparent abnormalities of early pregnancy to be the sole cause of diplegia in up to 16% of cases. It is tempting to suggest that in a proportion of premature patients threatened abortion may possibly have been an indication of foetal abnormality even at this early stage of /

of gestation.

Late pregnancy.

The most common disorders of the last trimester of pregnancy amongst diplegic patients were pre-eclamptic toxaemia and antepartum haemorrhage (Tables 105 to 106). It will be observed that pre-eclampsia was the most frequent abnormality of late pregnancy in mature patients, occurring in ten of the 40 (25%) about whom full details were known, and that antepartum haemorrhage was the most frequent complication of late pregnancy in the premature patients, occurring in 12 of the 34 patients (40%) about whom full details were known.

Pre-eclampsia was present in pregnancy in 16 of the 74 patients with diplegia in the series about whom full details were known (approximately 21%). It occurred in 25% of mature cases and in about 18% of the premature, a difference which is not statistically significant on the small numbers of the series. In two of the premature patients and in two of the mature patients the pre-eclampsia was mild; in two of the premature and one of the mature it was classified as severe. Antepartum haemorrhage occurred as a complication in two premature patients and in one mature patient. Pre-eclampsia was preceded by threatened abortion in one premature and in two mature cases (Tables 105 and 106).

Antepartum haemorrhage occurred in 16 patients in the series of 74 about whom full birth details were known (approximately 21%). Of the 34 patients who were prematurely born /

TABLE 105

Pre-eclampsia in the pregnancies resulting in the birth of diplegic patients

	Case No.	Mat. Age	No. of Preg.	Severity of pre-eclampsia	Other disorders of pregnancy	Labour	Dur. of labour hrs.mins.	Delivery	Drugs	Condition of child	Birth Wgt. lbs.	oz.	Placental Infarctions
MATURE	22	38	2	Moderate	-	Normal	23 35	Spont.vertex	Chlorof.	Poor. Apnoea	5	15	+
	57	38	1	Mild	Threatened abortion at 3 months.	Uterine inertia	16 0	Mid cavity forceps	Hyoscine Chlorof.	Poor. Apnoea	9	-	-
	59	42	2	Mild	-	Normal	17 15	Spont.vertex	Nitrous oxide	Good	5	13½	+
	118	18	1	Moderate	-	Medical induction	11 40	Spont.vertex	Chlorof.	Good	7	14	+
	126	32	1	Moderate	Antepartum haem.	Precipitate	minutes	Spont.vertex	-	"Quite good"	6	8	-
	131	23	1	Moderate	Attempted abortion. External version.	Dry labour	27 15	Low forceps	Trilene Ether	Good	6	15	+
	138	23	1	Mild pre-eclampsia	-	Normal	18 40	Spont.vertex	Pethedine	Good	8	5½	-
	141	36	1	Moderate	Vomiting for 1 mth.	Inertia. Foetal distress	103 30	Mid cavity forceps	Pentothal Cyclopro-pane	Poor. Apnoea	5	14	+
	147	31	1	Severe	Hydramnios	Artif.rupture membranes. cord prolapse.	72 -	Mid cavity forceps	Paralde-hyde.	Good	6	12	-
	198	24	1	Moderate	Hyperemesis	Medical induct. Foetal distress	6 40	Spont.vertex	Chlorof.	Good	6	9½	+
PREMATURE	57	38	1	Mild	Threatened abortion	Uterine inertia Maternal distress	16 -	Mid cavity forceps	Hyoscine Omnopon Chlorof.	Poor. Prolonged apnoea	5	4	-
	124	37	2	Moderate	-	Prolonged	72 -	Spont.vertex	Unknown	Fair. No apnoea	3	11	-
	155	30	4	Mild	Recurrent pyelitis. Antepartum haem.	Normal	6 35	Spont.vertex	Nitrous oxide	Poor. Apnoea	5	8	-
	156	21	1	Severe	Severe vomiting	Bougie induction. Inertia	37 25	Spont.vertex	"Luminal"	Normal premature	4	8½	+
	157	34	3	Moderate	Goitre. Anaemia. Placenta previa. Antepartum haem. Much morphia.	-	-	Caesarean section	Ether	Poor. apnoea	5	-	-
	186	35	2	Severe	Thyrotoxicosis	Precipitate	1 15	Spont.vertex	Normal	"Normal premature"	5	2½	+

TABLE 106

The birth histories of patients suffering from diplegia

Antepartum Haemorrhage

	Case No	Mat. Age	No. of Preg.	Maternal health	Pregnancy	Labour and Complications	Dur. of labour hrs.mins.	Delivery	Condition of child	Bth. Wgt. lbs.oz.	Placental Infarctions
MATURE	25	19	1	Normal	Antepartum haem. from placenta previa at 36 weeks.	Medical induction	2 -	Willetts forceps	Limp.Poor Apnoea.	5 15	+
	38	23	2	Good	Antepartum haem. at 18 28 & 39 weeks. External version.	Prolonged	48 -	Spont.vertex.	Poor. Apnoea	7 4	-
	42	28	5	Resp.infections.	Fall at 6½ mths.followed by antepartum haem.	Traverse lie.	24 -	Breech extraction	Poor. Apnoea	9 -	-
	126	32	1	Good	Moderately severe pre-eclampsia.Antepartum haem.at 34 wks.	Pituitrin given. Precipitate.	mins.	Spont.vertex.	Poor. Apnoea	6 8	-
	2	31	3	Good	Much vomiting. APH 3 days before delivery.	Prolonged	84 -	Spont.vertex.	Apnoea. Cyanosed. Incubated.	3 3	-
	3	34	2	Good	APH at 8 mths. immediately prior to labour.	Morphia and med. induction.	- 15	Precip.followed by forceps deliv. through narrow outlet.	Apnoea	4 15	-
	61	32	2	Healthy	Assaulted at 2 mths. Attempted abortion 3 & 4 mths. Antepartum haem. at 29 weeks.	Prolapsed cord	3 40	Assisted bfeech	Apnoea	3 5	+
	83	33	1	Normal	Haem. at premature onset labour at 28 weeks.	Foetal distress Morphia & pituitrin.	47 -	Fundal pressure	Fair	2 14	-
PREMATURE	152	34	2	Healthy		Artif.rupture memb. Pethedine	3 45	Breech extract.	Poor	3 11	+
	157	34	3	Goitre.Anaemia	Pre-eclampsia.Haem. at 27 & 32 weks. Much morphia.	Caesarean section	-	Caesarean sect.	Poor	5 -	-
	172	32	1	Normal	Threatened abort. 1st. & 2nd. mths. APH prior to delivery.	Prolonged labour. Contracted pelvis.	72 -	Spont.vertex	Poor	2 12	-
	174	23	3	Normal	Bleeding at 2½ & 7 mths. from placenta previa.	Internal version. Leg brought down.	24 -	Asisted breech	Very poor Marked apnoea	3 -	+
	181	30	3	Normal	Twin preg.Hypertension for 3 mths.Attempted abortion at 3½ mths.	Normal twin labour.	8 50	Spont.vertex. Foetus papyraceous.	V. poor Marked apnoea	2 8½	+
	13	22	2	Tuberculosis Ovariectomy	Antepart.haem. 30 wks.	Normal premature	5 45	Spont.vertex	Apnoea	3 8	-
	140	24	3	Ovariectomy	Vomiting 5 mth.Antepart.haem. 21,31,33 wks. Pethedine.	Normal premature	21 30	Spont.vertex	Poor	3 10	+
	155	30	4	Pyelitis	Pyelitis.Mild pre-eclampsia Antepart.haem.day before	Normal premature	6 35	Spont.vertex	Poor.apnoea	5 8	-

TABLE 107

Comparison of abnormalities of pregnancy which might be considered causes of premature birth in prematurely and maturely born patients.

	Premature diplegics			Mature diplegics		
	Number	%	Approx. %	Number	%	Approx. %
1. Twin pregnancy. Uncomplicated.	3	9	12	0	0	0
Complicated.	1	3		0	0	0
2. Singleton pregnancies.						
Pre-eclamptic toxaemia ... only	3	9	18	7	17	24.
" " with haemorrhage before or after 28th. week.	3	9		3	7	
Threatened abortion or haemorrhage before 28th. week only	2	6	33	2	5	12.
Before and after 28th. week.	4	12		1	2.4	
After the 28th. week only.	5	15		2	5	
Hydramnios	1	3		1	2.4	2.
Other						
Pyelitis only.	1	3	15	1	2.5	7.
Other infections.	2	6		1	2.5	
Fainting frequently.	1	3		0	0	
Trauma without haemorrhage.	1	3		1	2.4	
Foetal abnormality other than diplegia						
Pregnancy uncomplicated.	2	6	15	5	12	22
" complicated. ⁂	3	9		4	10	
No apparent complication of pregnancy. Foetus free of other malformations.	5	15		18	43	
Total number of pregnancies.	33	99		42	100	

⁂ Not an exclusive category and not included in total

similar history.

Of the total of 18 patients delivered prematurely in whom there was a history of abnormalities of late pregnancy, one had a history of threatened abortion previously (Case 140) and 14 had a history of later abnormal deliveries, of whom 5 had had abnormalities of early pregnancy in addition. Of the 16 mature patients with a history of abnormalities of the last trimester of pregnancy, in six this appeared to be the only abnormality of parturition, but in 10 labour or delivery was complicated and in two of these (Cases 38 and 131) there were additional abnormalities of early pregnancy.

Thus in only three premature patients (Cases 13, 186 and 155) and six mature patients (Cases 10, 22, 59, 118, 120 and 138) were abnormalities of the last trimester of pregnancy the only disorders of pregnancy, labour and delivery. In the remaining cases there were other complications of parturition.

In view of the fact that 40 of the 74 fully detailed patients had no abnormalities of late pregnancy, these cannot be regarded as being a constant necessary aetiological factor in diplegia. That these abnormalities occur more frequently in association with other abnormalities of pregnancy, labour and delivery than alone in the histories of diplegic patients makes it likely that if they are of aetiological significance they are more often contributory than directly causal. On the other hand the possibility that the association of abnormalities of pregnancy, labour and delivery and the occurrence /

occurrence of diplegia in the foetus may not be the result of cause and effect, but due to the presence of underlying factors causing both must be remembered.

Abnormalities of labour and delivery.

Nineteen of the 40 fully detailed mature patients (approximately 47%) had a history of abnormal labour or delivery, and 22 of the 34 prematurely born patients (approximately 65%) had a similar history. Thus a total of 41 of the 74 diplegic patients under consideration, or approximately 55%, had disorders of labour or delivery or both.

(a) Labour. In ten of the 19 mature patients with abnormalities of labour or delivery there was prolonged labour lasting more than 30 hours in all or including a second stage of more than 24 hours' duration (Table 108). A similar history was present in 5 of the 22 premature patients with abnormal labour or delivery. The prolonged labour was attributed to disproportion in one of the premature cases (Case 172), though on the face of it this seemed unlikely, and to malpresentation in one of the mature cases (Case 81). In 6 of the mature and one of the premature cases uterine inertia was blamed for the prolonged labour. In one of the mature cases (Case 131) there was dry labour. In two of the mature patients (Cases 38 and 147) and three of the prematures (Cases 2, 83 and 124) no cause for the prolonged labour was stated though inertia was probably the cause in the majority. There was evidence of foetal distress during labour in one of the premature cases (Case 83) and in two of the mature (Cases

TABLE 108

Abnormalities of labour in diplegic patients - Mature

Prolonged 2nd. stage labour

Case No.	Mat. Age	No. of Preg.	Pregnancy	Labour	Dur. of labour hrs,mins.	Delivery	Drugs	Condition of child.	Bth.Wgt. lbs.	oz.
38	23	2	Antepartum haemorrhage. External version.	Prolonged.	48 -	Spont.vertex	-	Poor. Apnoea	7	4
81	25	1	External version 4 times. Attempted abortion 3½ mths.	Traverse arrest.	53 -	Manual rotation. Difficult mid-cavity forceps.	Heroin Chlorof.	Normal	6	2½
131	23	1	Pre-eclampsia. External version 7 mths.	Dry labour.	27 15	Low forceps.	Chlorof.	Good	6	15
141	36	1	Vomiting for one mth. Mod. severe pre-eclampsia.	Prolonged. Uterine inertia. Foetal distress.	103 30	Mid cavity forceps.	Pentothal. Cyclopropane.	Poor. Apnoea	5	14
147	31	1	Severe pre-eclampsia and hydramnios.	Artificial rupture of the membranes.	72 -	Mid cavity forceps.	Paraldehyde.	Good	6	12
56	28	1	Premature rupture of membranes.	Uterine inertia.	120 -	Rotation of forceps extraction.	Chloral Chlorof.	Good	6	8
65	24	1	Normal.	Uterine inertia.	24+	High forceps delivery.	Chlorof.	Good	9	8
66	24	1	Vomiting. Obese mother.	Uterine inertia. Maternal and foetal distress.	120 -	Rotation and forceps deliv.	N_2O. Ether	Good	6	8
104	28	1	Normal.	Uterine inertia.	24*	Mid cavity forceps.	Chlorof.	Good	7	4
200	17	1	Normal.	Prolonged labour. Uterine inertia.	82 -	Spont.vertex.	-	Very poor	7	8
				Precipitate labour						
126	32	1	Pre-eclampsia and antepartum haemorrhage.	Pituitrin induction.	minutes	Spont.vertex	-	Normal	6	15
55	34	2	Normal.	Medical induction.	15	Spont.vertex	-	Normal	7	7
154	26	1	Normal	Precipitate.	minutes	Spont. vertex	-	Normal	7	0
34	36	4	Normal	Medical induction.	minutes	Spont. vertex	Morphia.	Poor. Apnoea	7	0
10	35	2	Normal	Ergot. Precipitate.	"about 2 hours.	Spont. vertex	None	Good	9	8

TABLE 109

Abnormalities of labour in diplegic patients - Premature

A Prolonged 2nd. stage labour

Case No.	Mat. Age	No. of Preg.	Pregnancy	Labour	Dur. of labour hrs.	mins.	Delivery	Drugs	Condition of child.	Bth.Wgt. lbs.	oz.
2	31	3	Much vomiting and haemorrhage for three days before delivery.	Prolonged.	84	-	Spont. vertex.	Trilene.	Apnoea. Cyanosed.	3	3
83	33	1	Haemorrhage at 28 weeks.	Prolonged and foetal distress.	47	-	By fundal pressure. Vertex.	Much morphia.	Fair. No apnoeic.	2	14
124	37	2	Moderately severe pre-eclampsia.	Prolonged. Much morphia and pituitrin.	72	-	Spont. vertex.	Unknown.	Fair.	3	8
156	21	1	Severe pre-eclampsia.	Bougie induction and inertia.	37	25	Spont. vertex.	Luminal.	Normal premature.	4	8½
172	32	1	Bleeding at 1 and 2 mths. of pregnancy.	Contracted pelvis. Prolonged labour.	72	-	Spont. vertex.	None.	Poor. Apnoea.	2	13

B Precipitate labour

3	34	2	Haemorrhage at 8 months.	Morphia. Medical induction. Precipitate, followed by low forceps for outlet contraction.	45		Low forceps.	Chlorof.	Moderately severe apnoea.	4	15
51	23	1	Much vomiting. Persistent fainting. Premature rupture of membranes.	Precipitate.	30		Spont. vertex.	Trilene.	Fair with apnoea.	4	-
163	33	3	External version 24 hrs. before delivery after several days pain.	Very rapid.	minutes		Spont. vertex.	None.	Normal premature.	4	-
53	42	4	Obese bronchitic mother.	Rapid.	1 hour		Spont. vertex.	Morphia.	Very poor, Apnoea.	2	8
77	16	1	Normal pregnancy.	Very rapid.	Few mins.		Spont. vertex.	None.	Cyanosed and feeble.	3	15
186	35	2	Thyrotoxic. Severe pre-eclampsia.	Precipitate.	75		Spont. vertex.	None.	Normal premature	5	2½

141 and 66). It will be noted that three of the mature patients (Cases 38, 141 and 200) were delivered in very poor condition, and that two of the premature patients were severely affected after birth (Cases 2 and 172). The condition of a further two was described as fair (Cases 83 and 124). To conclude that prolonged labour is a probable cause of diplegia in these patients, however, is not valid, for pregnancy was complicated in all five of the prematurely born cases and in five of the ten mature. Heavy sedation or anaesthesia was given in at least four of the cases, and in 8 of the mature patients instrumental delivery was required. Thus there were other potential causes of foetal damage in pregnancy and delivery in all of the histories with the exception of Case 200 in which prolonged labour and uterine inertia was the only apparent abnormality.

Precipitate labour lasting less than two hours occurred in 6 prematurely born (Tables 101,102) and in 5 maturely born patients (Tables 94,95). In all the premature cases except Cases 53 and 77 pregnancy had been complicated, and only two infants (Cases 163 and 186) were born in fair or good condition. Only one of the mature patients delivered precipitously had a history of abnormal pregnancy (Case 126), and only one infant was born in other than good condition (Case 34) in which much morphia had been given during labour. Thus of the total of 11 patients in whom there was a history of precipitate labour, only 5 were apparently free of other disorders of pregnancy, labour or delivery.

Thus /

TABLE 110

Forceps delivery in patients with diplegia

	Case No.	Mat. Age	No. of Preg.	Pregnancy	Labour	Dur. of labour Hrs.mins.	Delivery	Drugs	Condition of child	Bth.Wgt. lbs. oz.
	18	29	1	Normal.	Foetal distress.	3 30	High forceps.	Chlorof.	Poor. Apnoea.	5 12
	65	24	1	Normal.	Uterine inertia.	72 -	High forceps.	Chlorof.	Normal.	9 8
	66	24	1	Vomiting and obese mother.	Inertia. Persistent occipito posterior. Maternal and foetal distress.	12 -	Manual rotation. Forceps extraction.	N_2O. Ether.	Good.	6 8
	104	28	1	Normal.	Uterine inertia.	24 -	Mid cavity forceps.	Chlorof.	Quite good.	7 4
MATURE	25	19	1	Severe antepartum haemorrhage.	Surgical induction. Willetts forceps.	2 -	Vertex and Willetts Forceps.	Chlorof.	Limp and apnoea.	5 15
	57	37	1	Threatened abortion. Pre-eclampsia.	Uterine inertia.	16 -	Mid cavity forceps.	Hyoscine. Chlorof.	Facial palsy. Otherwise well.	6 4
	81	25	1	External version four times.	Prolonged. Traverse arrest.	53 -	Manual rotation. Difficult mid-cavity forceps.	Heroin.	Good.	6 2½
	131	23	1	Attempted abortion at 31 mths. Moderately severe pre-eclampsia. External version at 7 months.	Premature rupture of membranes. Dry labour.	27 15	Low forceps.	Trilene. Ether.	Good.	6 15
	141	36	1	Vomiting for one month. Pre-eclampsia.	Prolonged uterine inertia. Foetal distress.	103 30	Mid cavity forceps.	Pentathol. Cyclopropane.	Poor. Apnoea.	5 14
	147	31	1	Severe pre-eclampsia and hydramnios.	Artificial rupture of membranes. Prolonged.	72 -	Mid cavity forceps.	Paraldehyde.	Good.	6 12
	3	34	2	Haemorrhage at 8 months. Morphia and medical induction.	Precipitate followed by forceps delivery through small outlet.	- 15	Low forceps.	Chlorof.	Good.	4 15
PREMATURE	57	37	1	Threatened abortion at 3 mths. Mild pre-eclampsia.	Uterine inertia 2nd. stage delivery.	16 -	Mid cavity forceps.	General anaesthesia.	Prolonged apnoea. Feeble.	5 2

Thus it is impossible to regard precipitate delivery as more than one of a number of possible factors of aetiological importance in diplegia in the majority of cases in which it occurs, and the possibility of it being merely a coincidental finding has to be remembered.

(b) Delivery. Forceps delivery was required in 12 cases, 10 of the 40 mature (25%) including Case 25 in which Willett's forceps were used, and 2 of the 34 premature (approximately 6%) (Table 110). Since forceps are rarely applied without reason the finding that all the patients had other abnormalities of pregnancy, labour or delivery is hardly surprising. Six of the mature and both the premature patients had abnormalities of pregnancy together with abnormalities of labour, and the remaining 4 mature cases (Cases 18, 65, 66 and 104) had abnormalities of labour only. In view of these findings it is extremely difficult to ascribe to forceps delivery, per se, the occurrence of cerebral damage in the foetus. In fact, the presence of foetal distress in at least 3 patients (Cases 18, 66 and 141) prior to the application of forceps might be regarded as suggestive evidence that the infants had already suffered some damage.

One mature patient (Case 42) and 5 premature patients presented by the breech (Cases 158, 41, 63, 61 and 152). The first three of these were twins. In Case 174 internal version was performed and a leg brought down to stop the bleeding from a placenta praevia. All the above 7 patients were delivered by breech extraction, four in poor condition, one /

one in fair condition and two in good condition considering their birth weights. The fact that the incidence of breech extraction in the premature group is only 6 patients, including three twins, in the total of 34 prematurely born diplegic cases (approximately 17%) suggests that this mode of delivery cannot be a major direct cause of diplegia. An incidence of 10% for breech delivery amongst premature singleton births of multiparous mothers was found in an unselected sample of hospital births by Drillien (1947). Moreover, though all three twins were born after apparently uncomplicated pregnancies there were abnormalities of labour in three (Cases 61, 152 and 174).

In three premature cases there were other forms of abnormal delivery. In Case 157 Caesarean section was performed at 32 weeks' gestation after a pregnancy complicated by pre-eclamptic toxaemia and antepartum haemorrhages for which much morphia had been given. In Case 83 the infant was delivered by direct fundal pressure at an estimated 29 weeks' gestation when the heart sounds had ceased to be audible. Pregnancy had been complicated by a severe antepartum haemorrhage and labour had been prolonged. In Case 159 (the second born twin of Case 158) the child was delivered spontaneously in the occipito posterior position after a normal labour.

Neonatal apnoea and hypoxia.

It will be seen from Tables 112 and 113 that of the total of 74 patients with congenital diplegia about whom reasonably /

reasonably full birth details are known, 41 (approximately 55%) appeared to be normal immediately after delivery, and a total of 27 (36%) appeared to be normal, apart from prematurity in the premature group, during the neonatal period. These figures indicate that abnormal neonatal periods are by no means a constant finding in diplegic patients, and that neonatal apnoea cannot be incriminated as even a contributory factor in the majority of patients, especially those who are born maturely. (Table 111)

It will be observed, however, that whereas 15 of the 19 patients (approximately 79%) in whom pregnancy and delivery appeared to be normal showed no abnormality immediately after delivery, only 9 of the 24 (37.5%) in whom both pregnancy and delivery had been abnormal appeared to be healthy immediately after delivery. The total with only abnormal pregnancy who appeared to be normal immediately after delivery was 7 of the 15 patients (approximately 46%); with abnormal delivery it was 10 of 16 (approximately 63%). There is thus suggestive evidence that the occurrence of neonatal apnoea is more closely related to the presence or absence of abnormalities of pregnancy and delivery than to the presence or absence of diplegia.

When the groups of premature and mature infants are compared it is seen that a rather smaller proportion of the former than the latter were normal immediately after delivery (excluding size from consideration). As might be expected the proportion of premature infants with a normal subsequent course /

TABLE 111.

	Mature					Premature					
	Normal at birth		Abnormal at birth			Normal at birth		Abnormal at birth			
	Normal Neonatal Course	Abnormal Neonatal Course	Normal Neonatal Course	Abnormal Neonatal Course	Total	Normal Neonatal Course	Abnormal Neonatal Course	Normal Neonatal Course	Abnormal Neonatal Course	Total	Great Total
Pregnancy and delivery normal.	11	1	0	3	15	1	3	1	0	5	20
Pregnancy only abnormal.	4	1	0	3	8	2	0	4	1	7	15
Delivery only abnormal.	2	3	1	3	9	3	2	1	1	7	16
Pregnancy and delivery abnormal.	3	1	1	5	10	2	3	10	0	15	25
Total	20	6	2	14	42	8	8	16	2	34	76
	26		16		42	16		18		34	76

course was smaller than the proportion of matures with a normal subsequent neonatal course, 33% as compared to 53%. (Tables, 112, 113) Thus for diplegic, as for non-diplegic, infants low birth weight appears to be associated with a higher incidence of neonatal apnoea and of other complications in the neonatal period, than is found in the mature. On the other hand the incidence of neonatal apnoea and of complications in the neonatal period is higher in both groups than in random series of non-diplegic patients. This would appear to be consistent with the higher proportion of cases of diplegia with a history of abnormal pregnancy or delivery. On the evidence available from this study neonatal apnoea and complications cannot be regarded certainly as either a result or a cause of diplegia.

Further consideration of the histories of those patients with a history of apparently normal parturition.

A total of 13 of the 42 maturely born diplegic patients had no apparent abnormalities in pregnancy, labour or delivery and in four of the prematurely born there was no apparent abnormality apart from the premature delivery. The fact that possible or probable hypoxic, toxic or traumatic factors were not in evidence in such a high proportion of diplegic cases (24% of the total detailed of 74) makes it necessary to consider them further from the aetiological point of view. In particular they must be compared with the groups delivered after abnormal parturition to see if any possible hereditary or environmental factors of possible aetiological importance may be defined.

Comparison /

TABLE 112

Comparison of condition of patient immediately after birth and in the neonatal period by premature and mature birth

	Mature		Premature		Total	
	Number	%	Number	%	Number	%
Normal after birth. Normal neonatal period.	20	48	8	23.5	28	37
Normal after birth. Abnormal neonatal period.	6	15	8	23.5	14	19
Abnormal after birth. Normal neonatal period.	2	5	2	6	4	5
Abnormal after birth. Abnormal neonatal period.	14	32	16	47	30	30
	42	100	34	100	76	100

TABLE 113

Comparison of the proportions of normal and abnormal pregnancies and deliveries of prematurely and maturely born diplegic children and other pregnancies of their mothers.

	Premature group.		Mature group.	
	Patients. % of 33	Other Pregs. % of 69	Patients. % of 42	Other Pregs. % of 70
Normal pregnancies and deliveries.	15	46	35	76
Abnormal pregnancies and deliveries	85	54	65	24

TABLE 113

Comparison of postnatal conditions of the child by maturity and normality of pregnancy and labour or delivery

		Normal after birth							
		Normal neonatal period.		Abnormal neonatal period.		Normal neonatal period.		Abnormal neonatal period.	
		Number	Approx %	Number	Approx %	Number	Approx %	Number	Approx %
Pregnancy and labour or delivery normal.	Mature	11	2.7	1	2.3	0	0	3	7
	Prem.	3	9	1	3	0	0	1	3
Only pregnancy abnormal.	Mature	4	10	1	2.3	0	0	3	7
	Prem.	0	0	2	6	1	3	4	12
Only delivery abnormal.	Mature	2	5	3	7	1	2.5	3	7
	Prem.	2	6	3	9	1	3	1	3
Both pregnancy and labour or delivery abnormal.	Mature	3	7	1	2.3	1	2.3	5	12
	Prem.	3	9	2	6	0	0	10	29
Total	Mature	20	49	6	13.9	2	4.8	13	33
	Prem.	8	23.5	8	23.5	2	6	16	47

Comparison of Distribution by Social Class.

The group of patients without a history of abnormal pregnancy, labour or delivery was compared by Social Class with the group with a history of abnormal parturition (Table 114). It will be seen that there is a quite striking tendency for the former to have a much more "normal" distribution than the group with a history of abnormal parturition. Whereas there is a preponderance of the latter group in Social Classes IV and V the largest group of the cases with normal pregnancy, labour and delivery is in Social Class III. In spite of the small number of cases the difference between the groups is probably significant statistically (p being 0.02 approximately).

The patients with a history of possible or probable injury during pregnancy, labour or delivery show a distribution by social class very similar to that observed in congenital hemiplegia, in which a similar birth history is so frequently obtained. On the other hand the patients suffering from diplegia in whom there is no evidence suggesting injury during pregnancy, labour or delivery show a distribution more in keeping with that of all births in Counties of Cities (Registrar General for Scotland, 195).

Comparison of Birth History by Family Size.

No significant or striking difference could be found in the average sizes of families in the two groups nor in the number of other pregnancies to the mothers of prematurely and maturely born diplegic patients.

The /

The proportions of fathers and mothers suffering from neurological and mental abnormalities were also similar. Comparisons were also made between the proportions of abnormal relatives, siblings, stillbirths and neonatal and post-neonatal deaths. It was found that there was a greater proportion of mentally retarded, congenitally physically abnormal siblings and dead siblings in the group of cases in which patients were delivered after normal parturition than in that in which they were born after abnormal parturition. Unfortunately the difference is only of doubtful statistical significance (p being between 0.05 and 0.02). The difference was not due to the mothers with normal pregnancies, labours and deliveries with the patients having a higher incidence of abnormal other pregnancies. On the contrary the mothers giving birth to patients after normal parturition had a significantly higher proportion of other normal pregnancies (Table 116). It will be recalled that there was a greater proportion of premature births in the group in which patients were delivered after abnormal pregnancy, labour and delivery than in that in which they were delivered after normal pregnancy, labour and delivery. Since it has already been pointed out that mothers with prematurely born patients tended to have a higher incidence of other abnormal pregnancies than those giving birth to mature patients, these findings are not surprising. It does appear to indicate, however, that there may be a greater tendency for the mothers with patients delivered after normal birth to produce more abnormal children than those in

which /

TABLE 114

Distribution by social class of diplegic patients.

Social Class	I	II	III	IV	V
No abnormality of pregnancy or delivery.	1	2	9	2	3
		3	9	5	
Abnormal pregnancy, labour or delivery.	4	11	17	12	13
		15	17	25	

(p-.02)

TABLE 115

Patients with diplegia by birth weight and normality of pregnancy labour and delivery

	Normal pregnancy, Labour and delivery	Abnormal pregnancy, labour or delivery	All
Mature patients.	13	29	42
Premature patients.	4	28	32
Total	17	57	74

(p-.01)

TABLE 116

History of other pregnancies to mothers with diplegic children by normality of parturition

	Normal parturition	Abnormal parturition	All
Number of other pregnancies.	35	103	138
Number of other normal preg.	28	57	85
Total	63	160	

(p-.01)

TABLE 118

Siblings of diplegic patients by normality and normality or abnormality of parturition of the patients.

	Number of patients	Abnormal siblings Mentally	Physically	Dead siblings
No abnormality of parturition.	17	6	5	3
Abnormal parturition.	57	7	7	5
Total	74	13	12	8

TABLE

Patients with diplegia by maternal age and normality of parturition

Maternal age	Normal parturition	Abnormal parturition	All
15-25	6	21	27
25-35	5	31	36
35 plus	6	7	13
All	17	57	74

which patients were delivered as a result of complicated parturition. That the higher incidence of abnormal siblings is unlikely to be due to birth injury has already been stressed. It appears that in the group of mothers giving birth to diplegic patients as a result of apparently uncomplicated parturition there may be a considerable proportion in whom there is a tendency for developmental abnormalities to occur as a result of genetic factors or causes unknown.

Comparison by Maternal Age.

The groups were compared by maternal age. It will be observed (Table 117) that a much higher proportion of the mothers giving birth to patients after abnormal parturition are between 25 and 35 years of age than are those with patients born after uncomplicated pregnancy and delivery. A higher proportion of the mothers whose diplegic children were born after uncomplicated pregnancy and delivery were over the age of 35 years. In spite of the small numbers of patients these differences are statistically significant (p being less than 0.01). On the other hand the proportions of mothers below the age of 25 in the two groups are very similar.

The size of the difference in the distribution of the groups by maternal age is the more striking when it is remembered that the incidence of abnormal pregnancy, labour and delivery does increase with maternal age, especially over the age of 35. On the other hand the proportion of children showing congenital malformations increases with age even /

even more sharply and it is impossible to avoid the speculation that in this fact may lie part of the explanation of why the maternal age is higher in the group without a history of abnormal parturition.

If this speculation were correct, and it could be shown that older women tend to bear diplegic children (in the same way as mongols) more frequently than younger women, the difference in the social class distribution would also be explained, since there is a significantly higher maternal age in Social Classes I and II than in III, and in III than in IV and V.

Conclusions.

From the evidence which has been offered, certain tentative conclusions may be drawn. Abnormal pregnancy, labour and delivery occur in a high proportion of cases resulting in the birth of diplegic patients. These abnormalities are significantly more frequent in prematurely born patients than in maturely born patients, and when they occur they are commonly multiple. A higher proportion of patients delivered as a result of abnormal pregnancy, labour and delivery are abnormal in the neonatal period than are those born after uncomplicated parturition, but it is still impossible to differentiate between those in which birth injury occurred and those in which it did not with any certainty. (Tables 96 and 103 and 119)

Mothers of diplegic patients are older and relatively infertile compared to mothers in the general population, and they /

TABLE 119

The frequency of abnormal pregnancy and delivery amongst prematurely born and maturely born diplegic patients

	Number of matures	%	Number of prematures	%	Total	%
Pregnancy and delivery normal (except for fact of premature labour in prematures).	15	35	5	15	19	27
Pregnancy only abnormal.	8	20	7	20.5	15	20
Labour or delivery only abnormal.	9	22	7	20.5	16	21
Pregnancy and labour or delivery abnormal.	10	23	15	44	24	32
Total	42	100	34	100	74	100
Unknown.	4		1		5	
Normal at birth and in neonatal period.	20	48	8	23.5		
Normal at birth but complicated neonatal period.	6	15	8	23.5		
Abnormal at birth. Normal neonatal period.	2	5	2	6		
Abnormal at birth and neonatal period.	14	32	16	47		
Total	40	100	34	100		

they appear to have a period in decreased fertility before and after the delivery of the diplegic child compared to the times they are giving birth to older and younger siblings. This suggests that conditions in the womb for the successful creation of a foetus may not be present. In this regard the high incidence of abortion to the mothers of diplegic patients immediately before and after the pregnancies with the patients is of interest. It is possible that the poor physical health of the mother noted in a relatively high proportion of cases may be associated with this relative inability to childbear successfully. The high proportion of gynaecological conditions and menopausal women is of interest.

Mothers giving birth to premature patients have a higher incidence of other abnormal pregnancies, labours and deliveries, especially abortions, than those giving birth to mature patients. (Table 120) Similarly mothers with a history of abnormal parturition with the patients had a higher incidence of abnormal other pregnancies, labours and deliveries. Mothers with patients delivered after apparently uncomplicated birth tended to be older and of higher social class than those with a history of abnormal births with the patients. They tended to have more congenitally mentally and physically abnormal children than did those in which parturition was complicated. This tendency to have more abnormal children could not be attributed to birth injury occurring more frequently. It is tempting to suggest that these women may have a greater tendency to produce children with developmental abnormalities

either /

TABLE 120

Comparison of labours and deliveries of prematurely and maturely born diplegic patients

	Prematures		Matures	
	Number	%	Number	%
A. Twin pregnancies. Normal labours.	5	15	0	0
" Breech extraction.	3	9	0	0
" Persistent occipito posterior.	1	3	0	0
" Spontaneous with foetus papyraceous.	1	3	0	0
B. Singleton births.				
(1) Normal labours and deliveries.	12	35	23	57.5
(a) Abnormal labours. Normal deliv.	9	27	9	22.5
Precipitate labour.	5	15	5	12.5
Prolonged labour. Disproportion.	1	3	0	0
Uterine inertia.	1	3	1	2.5
Cause not stated.	2	6	1	2.5
Foetal distress without other complications.	0	0	2	5.0
(c) Abnormal labour and delivery.	5	15	10	25.0
Precipitate labour. Low forceps.	1	3	0	0
Prolonged labour. Fundal pressure.	1	3	0	0
Low forceps.	0	0	1	2.5
Mid cavity forceps.	1	3	6	15.0
High forceps.	0	0	1	2.5
Intrapartum haemorrhage. Willetts forceps.	0	0	1	2.5
* Foetal distress. Forceps.	0	0	3	7.5
Prolapsed cord. Breech.	1	3	0	0
Malpresentation. Version. Breech extraction.	1	3	1	2.5
(d) Normal labour. Delivery abnormal.	3	9	0	0
Breech extraction.	2	6	0	0
Caesarean Section.	1	3	0	0
Total cases	34	100	42	100.0

* not an exclusive category

For one of two reasons. There may be a Mendelian recessive gene with different modes of expression as suggested by Penrose. But it seems more likely that the same reasons which cause other developmental abnormalities to occur more frequently in the offspring of older mothers, also operate in diplegia. Genetic mutation may be very important in this connection.

It is possible to define two groups of mothers of diplegic patients, both relatively infertile. Firstly, there are younger mothers, often of Social Classes IV. and V. with a high prevalence of abnormal pregnancies and deliveries, of abortions and of prematurity, whose diplegic offspring are usually born after abnormal pregnancy, labour and delivery. Secondly, there are older mothers (a slight excess of whom are in Social Classes I. and II.) with a lower proportion of abnormal pregnancies and deliveries.

Acquired Diplegia.

There was one case of acquired diplegia in the series whose case-summary will be presented.

Case 26. Born 1947.

He was the youngest of four children born to healthy parents of rather limited intelligence. He was born spontaneously by the vertex at home after a labour of "a few hours", and a normal pregnancy.

He sat with support at the age of four months and crawled before the age of seven months. He stood with support at the age of ten months and was walking alone quite confidently at the age of 11 months. By this time he was able to say "Mummy" and "Daddy", though he had no other words.

At /

At the age of 11½ months he had an attack of twitching of the left side of his face which lasted some minutes without accompanying loss of consciousness or other signs of illness. Because of uncontrollable crying and a recurrence of twitching, this time in the right face and upper limbs, he was taken to hospital where his temperature was found to be 102.4°F., but apart from intermittent twitching of the right side even during sedated sleep neurological examination gave normal findings. His white blood count was 8,400. A lumbar puncture gave normal findings. Systemic penicillin was begun in large dosage but fever persisted; there was twitching of the left face and occasionally of the right side of the face on the following day, and he seemed less aware of his environment. In the next week fever persisted and generalised convulsions occurred frequently and in addition occasional attacks of focal twitching of one side of his face, or one leg or one arm and a leg would occur. He moved much less in bed and gradually assumed a position of decerebrate rigidity, the right side showing more extension in rigidity than the left. He gradually recovered from this state and began to move all the limbs voluntarily again in the following week, but all movements appeared to be limited and clumsy, and the tendency to assume a position of opisthotonos persisted. He remained much less aware of his environment than before admission. He was transferred to a neurosurgical ward where a lumbar puncture revealed normal findings, except for a large gold curve of 3432000000 on the 18th day of illness. An air encephalogram showed /

showed a considerable degree of internal hydrocephalus without any lateral shift. He was thought to be suffering from thrombosis of the deep cerebral veins. He was discharged three weeks after admission, by which time his position of extension in all limbs had changed to one of predominant flexion in the upper limbs and extension rigidity in the lower.

Gradually, after his return home, his awareness of the environment improved and he began to follow objects with his eyes about three months after the onset of his illness. Eighteen months after the onset of his illness he was able to sit and began to use his left hand for holding and reaching for toys. He began to crawl shortly before the age of five but was still incontinent at that time.

Examination.

He was examined at the age of five and a half years. He was a rather undersized grossly defective child, drooling and unable to stand without support or talk intelligibly apart from a few phrases like "Go away". He was interested in his environment to a limited extent. His vision still appeared to be defective but his hearing was keen. He could respond to simple commands. He could stand with support. Head circumference was 19".

He appeared to have a right hemianopia. The optic discs were pale but otherwise the fundi were normal. There was right facial paresis. Swallowing was impaired. The right side of the tongue appeared somewhat atrophic.

His sitting balance was good but as soon as he was placed on his feet or held upright his feet went into equinus and the legs scissored. A position of opisthotonos was easily elicited by extending his head abruptly or putting pressure on the soles of his feet.

He could reach and handle objects clumsily with the left hand, but the right hand was held clenched and contracted, the arm generally being held in a flexed pronated position as in hemiplegia. Movements of his limbs were confined to shoulder and elbow movements which were full but weak. Supination was impossible. The right leg also moved poorly.

There was a generalised increase of tone in all limbs of mixed spastic and rigid type, spasticity being more evident on the right and in the arms than on the left and in the legs. The plantar responses were flexor. All the tendon jerks were increased, but much more markedly on the right than the left. Sensory findings were untestable.

During the examination he had two attacks lasting between 90 seconds and three minutes, in which there was slight tremor of all limbs, followed rather abruptly by the assumption of the decerebrate position and gross impairment of consciousness.

He was considered to show a picture of mixed rigid and spastic diplegia, probably the result of extensive cerebral destruction due to cerebral venous thrombosis of unknown cause. Occasional similar cases following extensive cerebral venous thrombosis have been recorded (Ford, 1952).

CHAPTER 7d

THE CLINICAL FINDINGS IN 79 CASES OF DIPLEGIA

The Course of Diplegia.

The majority of cases of diplegia have their origin before or at the time of birth, but it is usual for there to be a delay of some months in the appearance of the stage of diplegia marked by rigidity or spasticity. The majority of cases are only recognised by doctors and clinics when this final stage of spasticity or rigidity has appeared.

The change from an infant of a few weeks old, who shows little demonstrable abnormality on neurological examination, to the child of 2 or 3 years old with flexion contractures is a gradual one. But the sequence of changes that occur in the neurological manifestations marking the child's progress to this crippled state is remarkably constant. For the purposes of description three stages may be recognised in the gradual development of this final picture of spasticity with contracture of the limbs; the hypotonic stage, the dystonic stage and a third stage in which rigidity and spasticity are present together in varying degree in different patients.

The Hypotonic Stage.

In the first few weeks after birth the majority of children with diplegia show little obvious abnormality related to their neurological disorder. From the mothers or guardians of 31 patients in the present series, however, a history was obtained that suggested that the infants were hypotonic in the first weeks after delivery. "His head flopped all over the /

the place", "He felt like a half filled pillow for weeks and weeks" were typical comments. At the same time a high proportion of patients were noted to show marked poverty of movement. The child might retain the same position in which he had been placed when laid down in his cot for many hours. "He was so still you wouldn't think he had breathed all night" said one mother.

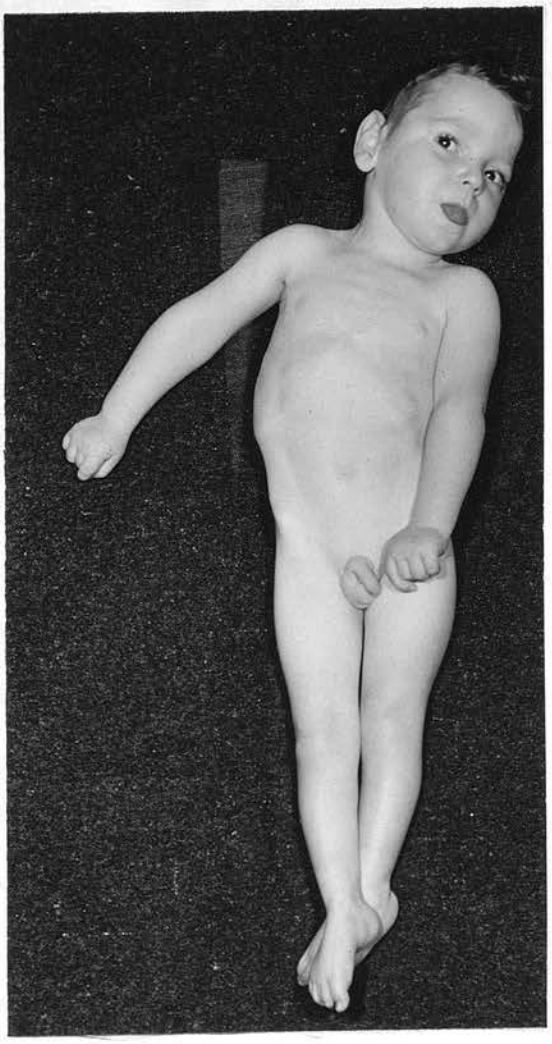
Twelve children have now been seen in this state, of whom only one is in the present series, the remaining cases living outside Edinburgh or being seen after the survey was completed. Poverty of movement is the striking feature and is especially marked in the trunk and proximal parts of the limbs. Apart from small range rather feeble finger movements, the patients were very still. On passive movement of the limbs well marked hypotonia is evident. In 11 of the 12 children the Moro and grasp reflexes were very marked and the tonic reflexes less marked than normal rather than exaggerated. The duration of the hypotonic stage varied from six weeks to 15 months in the patients in the series but has been observed in a child as old as 17 months. In 28 of the 31 patients in the series it appeared to last from between six weeks and six months. In general, the longer it lasted, the more severely handicapped was the child.

The Dystonic Stage.

At about the time the diplegic child first holds up the head a change occurs in the picture of little apparent abnormality. The mother finds that when she changes the child's /

Figure 4.

Posture in the dystonic stage of diplegia.



A severely affected diplegic patient showing the characteristic posture found in the dystonic stage. The position of the trunk and limbs is very similar to that found in opisthotonos and is most readily produced by sudden extension of the patient's head.

child's position abruptly, especially when dressing or bathing him, he suddenly becomes stiff in her arms. The neck and back are extended. The upper limbs are adducted and internally rotated at the shoulders, extended at the elbows, pronated in the forearms and semiflexed at the wrist and flexed at the fingers. The lower limbs are extended, adducted and somewhat internally rotated at the hips, extended at the knees, plantar flexed at the ankles and flexed at the toes. The position is essentially one of opisthotonos and is achieved by a sudden dystonic movement of the trunk and neck associated with extensor movements of the limbs. The position is maintained for only a few seconds, during which time the tone of the muscles is very greatly increased and the whole child is rigid. Sometimes there appears to be a slight impairment of consciousness during the attacks but it is never lost. Initially the attacks occur only once or twice a day, but over the course of one or two weeks their frequency increases. They become more easily produced and may occur as often as 20 or 30 times a day, greatly distressing the child. Frequently dystonic attacks are diagnosed as being epileptiform but they respond to none of the commonly used anticonvulsive drugs.

On examination at the height of the dystonic stage, it is usually found that the child is beginning to hold up the head, but is still unable to hold it steadily. He makes little attempt to reach for objects and poverty of voluntary movement is still evident. The tone of the limbs remains hypotonic at rest. The grasp reflexes are brisk. With care not to extend /

Figure 5 .Postures and reflexes in severe dystonic diplegia. Child aged 8 months.Resting neonatal position of semiflexion.(a) Moro reflex.(b).

a.

b.

Figure 6 .(Continued).Postures and reflexes in severe dystonic diplegia.Child aged 8 months.Reflex walking,(a). Sucking reflex,(b).Symmetrical neck reflex in erect position showing scissoring of lower limbs,(c).Tonic neck reflex,(d). Grasp reflex,(e).

a.

b.

c.

d.

e.

extend the head suddenly the Moro responses may be demonstrated in most of the patients. When the child is held vertically there is rigid increase of tone in the limbs. This is more marked in the lower than in the upper limbs. The lower limbs show a position of extension at the knees and plantar flexion at the ankles. The hips are adducted, slightly internally rotated and may show a position of full extension or of a few degrees of flexion. The legs may scissor. When the child's head is briskly extended, or he is suddenly rotated into the horizontal position from the vertical, supported by a hand in the small of the back, there is a sudden gross increase of tone in the trunk and limbs, associated with a quick, writhing, dystonic movement of the trunk. The limbs extend and a position of opisthotonos is assumed for a few seconds. This position corresponds exactly with that described by the mothers as resulting from changes in the position of the child when she handles him.

After a variable period the dystonic attacks become less easily produced and less frequent. In the majority of patients the generalized dystonic attacks are present for between two and six months, but in others the period may exceed one year. As the attacks begin to be less frequent the mother notices for the first time that the limbs are constantly rigid. This is first evident in the thigh or calf muscles and she may find that she has to use force to separate the thighs when placing the napkin, or even pass it between them on the end of a pencil. When she washes the lower /

lower limbs they feel stiff and unyielding. In the course of a few weeks the rigidity becomes increasingly evident and spreads to involve the upper limbs in triplegic and tetraplegic cases.

On examination during the period when dystonic attacks are waning a constant muscular rigidity is found, which is more severe in the lower limbs than in the upper. In the majority of patients sudden extension of the head still gives rise to a typical dystonic attack. In a few patients, however, it is necessary to extend the head and put pressure on the soles of the feet before dystonia is produced. When the child is made to exert himself, or is distressed, the affected limbs tend to assume extensor positions even if true dystonic movements of the trunk have ceased to occur.

During the survey six patients were classified as being in the dystonic stage of diplegia. Another seven with dystonia have been seen who are not included in the survey. In all, the essentially opisthotonic position described above could be produced by suddenly extending the head and putting pressure on the soles of the feet. Of the 79 children with diplegia in the survey, 39 had evidence of having had dystonic attacks, either in the history or on examination. It was noted that a history of dystonia was found much less frequently in older children than in younger and it is probable that if mothers' memories had been longer a greater number of patients who had had dystonic attacks would have been ascertained.

<u>The phase of Rigidity</u>.

The /

The Phase of Rigidity.

The third stage of diplegia is conveniently divided into two phases for the purposes of description, first the phase of rigidity and secondly the phase of spasticity.

As has been described, the phase of rigidity emerges gradually from the stage of dystonia and for a period, as rigidity increases in extent and severity, dystonic attacks persist. Somewhat arbitrarily the phase of rigidity may be said to begin, for purposes of classification, when generalised dystonic attacks involving the neck, trunk and limbs cease to occur as a result of changes in the child's position. This is usually about the time he attempts to sit with support with the knees flexed and is beginning to use the upper limbs in order to reach for objects. The parents notice that the attempts to grasp are clumsy and that the child appears to hold toys at arm's length with the affected upper limbs extended. In the majority of the patients in the series generalised dystonic attacks ceased between the ages of 5 and 12 months.

On examination children with predominant rigidity show clumsiness of voluntary movements. Manipulations with the hands are immature in type, all the fingers being used together as a unit opposed to the palms. The limbs tend to be held in positions of mild extension at rest, but this tendency is greatly increased by exertion, the performance of voluntary movements, extension of the head or pressure on the soles of the feet, especially when the child is held erect.

When /

Figure 7 .

Typical supported sitting and standing postures in early rigid/spastic diplegia. There is continuous antigravity hypertonus which is accentuated by the child being placed in the erect position with the head extended and pressure on the soles of the feet. The "decerebrate positions" of the arms are well shown. Only supported sitting is possible owing to the tendency to extensor thrust.

When an affected upper limb is moved voluntarily it tends to adduct at the shoulder, extend at the elbow, pronate at the forearm and flex at the wrist and fingers. This is the reason for the impression that the child is holding objects at arm's length. If the contralateral limb is also involved it too will show a tendency to attain a similar position to the operating limb and the lower limbs also extend. Thus associated movements of extension are present in all the other limbs when one is used and in all the limbs involved when the child exerts himself by straining or attempting to change his position. These associated movements are similar to those observed in the limbs during the earlier dystonic attacks. In the latter, however, the neck and trunk also took part and the involuntary movement was more rapid, more extensive and more powerful. In the stage of rigidity the involuntary postural activity appears to become more inhibited. Nevertheless it is found that the manoeuvres which were most potent in causing dystonic attacks at the earlier stage are also those which produce associated movements of limb extension most readily. When the child is held in the vertical position great rigidity of the lower limbs is evident and they are held scissored and either extended at the hips or in a few degrees of flexion. The knees are extended, the feet plantar flexed. The rigid increase of tone in the upper limbs is less accentuated and their positions may not be markedly constrained. If the feet are placed on the floor, however, or pressure is applied to the soles of the feet, the involved upper /

Figure 8.

Postures in the rigid/spastic stage of diplegia.

A moderately severely affected diplegic boy with well marked rigidity and some overlying spasticity developing, especially in the upper limbs. Antigravity tonus is increased by placing him in the erect position, extending the head or putting pressure on the soles of the feet. The lack of lumbar lordosis due to tightness of hamstrings when sitting is well shown.

Figure 9.

Postures in the rigid/spastic stage of diplegia. Continued.

Another moderately severely affected diplegic patient showing a little more spasticity than the one in the previous figure. Extension of the head, pressure on the soles of the feet and placing him in the erect position still results in a marked increase of antigravity tonus.

few patients but it is never brisk and in the majority it disappears completely during the stage of rigidity.

The Spastic Phase.

This phase is apparent only after rigidity of the muscles has been present for some weeks or months. In some spasticity appears only after the child has shown predominant rigidity for many years. Its onset is insidious and is usually apparent about the time the child is beginning to use the thumb and fingers a little and has learnt to sit without support with the knees flexed. Usually attempts to pull himself into the erect position follow shortly after this time.

The first evidence that spasticity has made its appearance is commonly a change in the nature of the associated movements of the limbs which results from voluntary activity or exertion. Over a period of a few weeks the parents notice that instead of being straight the upper limbs tend to be flexed and that all movements are less clumsy and jerky, though they may be slower than before. A grandmother noted, "His arms used to look as if they'd been put on the wrong way round but then they became all bent up". The same tendency to flexor positions rather than extensor positions becomes apparent in the lower limbs as in the upper. Unless the child is encouraged in all activities at the stage of early spasticity, especially standing and walking with support, flexion contractures rendering movement impossible can ensue in a few weeks. One nurse in an institution for the mentally defective summarised /

Figure 10 .

Characteristic postures in the late rigid/spastic stage of diplegia. There is considerable spasticity in all four limbs overlying rigidity, and flexion is beginning to predominate over extensor positions, in the limbs. (Case 81)

summarised the position when talking of a bedridden tetraplegic patient, "First he was too daft to move and now he can't".

On examination the findings in patients showing spastic increase of tone in the limbs depend very much on the relative severity of rigidity and superimposed spasticity in the individual patient. In patients who remain rigid, associated movements will remain extensor in type, and in patients who show early spasticity flexor associated movements make an early appearance. In the majority of diplegic patients, however, there is a transitional period during which spasticity is increasing in the limbs and the associated movements are neither typically extensor nor typically flexor.

By the time spasticity is well marked in the limbs, not only are the associated movements predominantly flexor but there is also a tendency for the child to hold the limbs in flexed positions at rest. These positions become exaggerated when the child exerts himself. Thus when he walks or attempts to walk there is flexion of the fingers, wrist and elbows, adduction of the thumbs, pronation of the forearms, and adduction and internal rotation of the shoulders in the affected upper limbs. An increase of flexion at the hips and knees, adduction of the thighs and plantar flexion also become evident. The typical walking position of the tetraplegic child results. The legs are scissored, the knees are semiflexed and the heels are not placed to the ground. The child appears to be leaning forwards with the upper limbs flexed /

flexed across the chest. When a spastic upper limb is used for grasping, athetosis is sometimes evident in the fingers and wrist, though this is not usually marked. Both in the operating and contralateral upper limb the flexor associated movements tend to be most marked at the distal joints in contrast to the extensor associated movements of the rigid phase, which are most marked in the proximal parts of the limbs.

The tonic neck reflex is very marked in the majority of patients in the spastic phase and both flexor and extensor positions of the upper limbs are readily demonstrable in triplegic and tetraplegic cases. The Moro response is absent and the grasp reflex present only occasionally. The tone of the limbs, and especially of the lower limbs, is a mixture of rigidity and spasticity, which gives rise to a very typical feel when they are moved passively. Stretch responses may be demonstrated in the muscles of the affected limbs, first and most easily in the adductors of the thighs and in the calf muscles. The tendon jerks are exaggerated, always to a greater extent in the lower limbs than the upper, and ankle and patellar clonus is frequent. The Hoffmann responses are frequently positive. The plantar responses are extensor bilaterally.

Once spasticity is evident, contractures in flexion are very liable to occur in the affected limbs. Contractures give rise to an increase of rigidity which is accentuated as the limbs are moved passively from the flexed to the extended position, /

Figure 11.

Postures in the rigid/spastic stage of diplegia.

Compared to previous illustrations the postures shown by this severely affected diplegic child are more consistently flexor. Spasticity is more prominent though underlying rigidity is still very evident. He shows a later stage in the evolution of the clinical picture of diplegia.

position, while the earlier rigidity of the limbs tended to be constant throughout the range of passive movement. The contracture is most likely to be severe in the lower limbs in diplegic patients, and it results, when severe, in a great diminution of the stretch reflexes and the tendon jerks. In those all too numerous patients in whom repeated tendon lengthenings, muscle transplants and joint fixations have been performed, the true neurological findings may be further concealed.

The Relationship of the Stages of Diplegia to Each Other.

The importance of recognizing that the findings in diplegia change in a regular manner as the condition develops is that this leads not only to earlier diagnosis, but also to better understanding of some of the variations which are encountered in the final clinical picture. The stages of diplegia mark definite milestones in the development of the child's motor control. Thus a child who shows dystonic attacks at the age of 4 years and is, therefore, in the stage of dystonia, must be regarded as showing a very immature motor control. On the other hand, a child in the phase of spasticity has passed through all the stages of diplegia and his neuromuscular mechanisms may be regarded as being more mature, though still disordered. Unless severely affected by contracture, he will be capable of much more in the way of controlled voluntary movement of the limbs than any patient in the stage of dystonia.

The /

The findings in the different limbs of patients with diplegia are not always those of the same stage. Thus it is quite common, even in children aged 10 or 12, to find that one limb is predominantly rigid while the other limbs are predominantly spastic. The child who holds one arm behind his back in the extended position in order to limit the range of its involuntary postural movements while walking and holds the other like a hemiplegic arm across his chest is not uncommon. Almost invariably the spastic limb is the more useful, unless contracture is severe.

It must be stressed that the stages of diplegia are determined by the relative maturity of the child's nervous system much more than by age. Thus one child was seen at the stage of hypotonia at the age of 2 years and one showed spastic diplegia at the age of 10 months. Nevertheless it is of some value to indicate the ages at which the majority of patients show the different stages. Of the 79 children in the present series, a history suggestive of hypotonia was elicited in 31. In 20 of these this stage lasted less than six months, and in 11 less than four months from the time of birth. The dystonic stage was noted in 39 patients. In 29 of these, it was evident before the age of 6 months and in eight between the ages of 6 months and one year. Only two patients gave a history which suggested that the dystonia had begun over the age of one year. The duration of the dystonic stage was unknown in four patients who were still showing dystonic attacks when the survey closed. In the remaining

TABLE 121

The age at the onset of dystonic attacks and their duration in 79 patients with diplegia

Age	Before 2 months of age.	Over 2 months Under 4 months	Over 4 months Under 6 months	Over 6 months Under 1 year	Over 1 year
Duration					
Less than one month.	0	0	1	0	0
More than one month, less than 2 months.	0	0	0	0	0
More than 2 months, less than 6 months.	0	5	3	1	0
More than 6 months, under one year.	0	4	3	1	0
Over one year, under 2 years.	0	2	1	0	0
More than 2 years.	2	0	0	1	0
Present at time of examination.	1	0	3	0	2
Totals	3	11	11	3	2

35 patients it lasted less than six months in 19 patients and between six months and one year in 11. In five patients the duration of the stage of dystonia was over one year. The age at which rigidity was first evident varied from 10 weeks to three years. In about half of the 72 rigid or spastic patients in the survey, no history of a previous dystonic stage was elicited, though that is not to say that it had not been present. (Table 121)

As has been noted, a few patients showed persistent rigidity throughout their lives without spasticity being superimposed upon this stage. A small number of patients appeared to develop spasticity at a relatively early age without a history of either previous dystonia or rigidity. The majority of patients, however, showed the sequence of stages - hypotonic, dystonic, rigid and spastic - which has been described. It was therefore much more usual to encounter cases showing mixed rigid and spastic increase of tone in the limbs than cases showing only rigidity or spasticity.

The age at which spasticity was first evident was even more variable than the age at which dystonia or rigidity first appeared. It was unusual to find spasticity before the age of 8 months, and in the majority of patients it made its first appearance between the ages of 10 and 18 months. On the other hand, the speed with which contracture could cripple the child with spastic diplegia was remarkable. One child was unable to extend either elbow beyond the right angle or pronate either forearm by the age of 18 months, though spasticity had

made /

made its appearance only at the age of 11 months so far as could be ascertained from the history.

The Severity of Diplegia.

Diplegic patients were classified for convenience into categories of paraplegia, triplegia and tetraplegia, according to whether useful function of the legs only, the legs and one arm, or all the limbs was impaired. The categories were based on a study of functional impairment rather than on neurological findings, however. In all but three cases labelled paraplegic, definite neurological abnormality of the upper limbs was apparent on careful examination. Impairment of the co-ordination of fine and rapid finger movements, impaired power of finger and wrist extension and of supination of the forearm were usually present. From the functional point of view, however, these findings were of relatively little importance in limiting the usefulness of the upper limbs.

Similarly cases called triplegic were all tetraplegic from the neurological point of view, but since the usefulness of only three limbs was significantly impaired, they were classified as triplegic from the functional point of view.

The degree of impairment of function varied considerably, even in those with paralysis of similar extent. Thus paraplegic cases might be completely unable to walk or their gaits might be only slightly abnormal. It was considered to indicate the severity of the paresis, as well as its extent, as discussed in the section on classification.

The /

TABLE 122

The severity of the paresis in 79 cases of diplegia in childhood

	Mild	Moderately severe	Severe	Totals
Stage of diplegia				
Hypotonic	0	1	0	1
Dystonic	0	2	4	6
Rigid or spastic	12	33	27	72
	12	36	31	79
Distribution of paresis				
Paraplegic	9	13	7	29
Triplegic	4	14	5	23
Tetraplegic	0	8	19	27
	13	35	31	79

The numbers of patients by distribution of the paresis encountered in the survey are indicated in Table 122 . It will be observed that the numbers in the categories of paraplegic, triplegic and tetraplegic paresis are roughly similar, but that the severity of the limb involvement tends to increase with the numbers of limbs involved. In tetraplegic patients it is more easily possible to observe the gradual change from dystonic to rigid and spastic stages and to account for some of their findings on the basis of the persistence of some of the dystonic phenomena into the rigid phase.

The Sex Distribution and Extent of the Paresis in the Diplegic Patients.

Thirty of the diplegic patients were female and 49 were male. The differences in the sex distribution in paraplegia, triplegia or tetraplegia were not significant.

It will be noted that there was an approximately equal number of patients showing paraplegic, triplegic and tetraplegic paresis.

These figures are roughly similar to those of Freud and the majority of modern series. Freud quoted a series of 69 cases of which 30 were of paraplegic distribution and 39 triplegic or tetraplegic. The only difference between this series and most modern series is that the latter tend to include a rather higher proportion of triplegic cases (Hellebrandt, 1951; Asher and Schonell, 1950). Obviously differences in the criteria used in classification and the nature of the cases included in the various series could

account /

account for these relatively small variations in the relative frequency of each form of paresis.

Cases in the Hypotonic Stage.

Since these patients in the stage of hypotonia are generally thought to be normal unless there are other congenital abnormalities, epilepsy, or the hypotonic stage is very prolonged, it is not really surprising that only one patient was seen who was an example of it. In the other cases at least a few dystonic attacks had occurred and the patients were therefore classified as being in the dystonic stage.

The exception was a very unusual form of diplegia of which I have only seen three examples in over 2000 cases of cerebral palsy in eight years. He was persistently hypotonic until the age of $2\frac{1}{2}$ and had never shown any evidence of dystonia. When first seen he was unable to sit with support even, but could support the microcephalic head. He was very mentally defective. In the next six months his limbs first showed very early spasticity and he learnt to sit with, and then without, support, and to use his hands, though rather clumsily. In his case the dystonic and rigid phases of the condition were missing. It was interesting that he had had repeated respiratory infections in infancy and had had feeding difficulty, as did similar mentally defective patients described by Yannet (1946).

Cases in the Dystonic Stage.

Six cases were in the dystonic stage of diplegia when they were examined and it was possible to observe the transition /

Case 13.

Severe dystonic tetraplegia, mental defect and retrolental fibroplasia.

A boy, born in 1952, the second of two children born to mother aged 22. Both parents tuberculous. All the family right handed. The first child was born at term after normal pregnancy and delivery, and is well.

In her second pregnancy the mother was extremely worried because her husband was ill and unemployed. She felt tired and had recurrent colds. Nine weeks before term she had a profuse vaginal haemorrhage. She was admitted to hospital and the child was delivered spontaneously under chloroform analgesia after a labour of 5¾ hours. The child was cyanosed at birth and would not breathe, but responded to oxygen and injections in a matter of minutes. The weight was 3 lb. 8 oz. The placenta was pale and weighed 15 oz.

The child was placed in an incubator and was in continuous oxygen for three weeks. At the end of this time the child developed a respiratory infection which necessitated penicillin and later blood transfusion. He was discharged at the age of 11 weeks when the weight was 6 lb. 8½ oz. It was noted at the time of discharge that the child did not appear to be seeing and that there was persistent nasal catarrh.

At the age of 15 weeks the child was admitted to hospital with pneumonia and in the first year of life received antibiotics in hospital and at home on seven occasions on account of otitis media, respiratory infections and bronchopneumonia. There was persistent chronic nasal catarrh.

When the child was discharged the mother noted that he felt "floppy" compared to her first child. He fed slowly, was windy, and at the age of 6 months the mother suspected he was backward because he was not attempting to hold up the head and was not looking about. He held up the head at the age of 8½ months, at which time bilateral retrolental fibroplasia was diagnosed. At the age of 10 months he was still not using the hands to play with toys, even when they were placed in them. The mother noted that when she washed him he frequently became stiff in her arms with the back arched, the head and legs thrown backwards, the forearms pronated, elbows extended and fingers semi-flexed. These dystonic attacks became more frequent and occurred on less stimulus, so that at the age of one year they were occurring many times a day whenever the child was handled. They were momentary only and the mother did not think there was any impairment of consciousness during them. The attacks persisted until the age of two, but from the age of about 20 months they gradually became less frequent and occurred only when the child was bathed and not on handling unless this was vigorous. At the same time his legs began to feel stiff.

At /

At the age of two years the child was able to sit with support but had no independent sitting balance, no appreciation of speech, showed no recognition of its mother and appeared to be able to see only the difference between light and darkness. He appeared to be unable to swallow solids. Many exacerbations of his chronic catarrh had occurred with fever.

Examination. Length 28½". H.C. 18". Fontanelle still ½" patent at the age of two years. Occasionally smiles, but otherwise very passive child resenting handling but showing no interest in his surroundings. Grunts and cries but no words. Much drooling. Able to hold up the head and sit with support. Moving the hands little, but the left more than the right. Severe bilateral retrolental fibroplasia. Right pupil irregular. Both pupils reacted slowly to light. Bilateral abducent paresis. Slight facial asymmetry due to right sided leg, on movement. Swallowing appeared to be infrequent.

There was weakness, stiffness and great inco-ordination of all limbs. This was very marked in the legs and the right arm but less marked in the left arm. There were no contractures, passive movements being full except for slight limitation of dorsiflexion of the feet bilaterally. There was a very severe increase of tone in the legs which were rigid, and a slightly less severe increase in the right arm. The increase in the left arm was moderate. Stretch responses were present in all muscle groups in all the limbs. The biceps, triceps, supinator, knee and ankle jerks were much increased in all limbs, rather more brisk in the right arm than the left. The Babinski signs were bilaterally present. The hands were very soft and the fingers were grossly hyperextensible, as were the toes. Postural reactions were marked, and interesting. When attempting to sit with support the child showed immediate flexion of the legs at the hips and knees and extension of the elbow with adduction and slight flexion at the shoulders, so that the arms were held rigidly and straight by the sides. When in this position the hands were tightly clasped. When the child was held in the horizontal position the arms were extended, as when sitting up, but the legs were also fully extended. When the child was held under the arm pits the legs were extended and scissored. But at the same time the arms were adducted at the shoulders, flexed at the elbows, wrists and fingers and pronated at the forearms. The tonic neck reflexes were present bilaterally, and the Moro response could be elicited inconstantly.

Sensory findings were untestable but light touch and pin prick were appreciated bilaterally. The hands and feet were cold to the touch.

Case 138.

A case of severe triplegia seen in the short dystonic phase.

An illegitimate male child, born in 1952 to healthy mother and father. Mother was aged 23 at the time of delivery. The patient was her first child.

She was well during pregnancy except that she was very large in the last 4 or 5 months of pregnancy on account of hydramnios. The labour was normal and lasted 18 hours. It occurred at term. The child was delivered spontaneously under nitrous oxide anaesthesia and appeared to be normal, crying at once. The weight was 8 lb. 5½ oz. The child was discharged from hospital to a nursery for the purposes of adoption. He seemed rather floppy for the first three weeks but fed well and no other abnormality was noted.

Between the age of 3 and 4 weeks he began to have spasms in which he suddenly went stiff, threw the head back and showed generalised rigidity and extension of the limbs. The attacks were momentary only and initially occurred most frequently when he was being bathed or washed. In the ensuing weeks he showed the attacks more frequently and at the same time they became more easily produced by less marked stimulation, especially feeding.

Examination at the age of 6 weeks. The child was alert and was making baby noises of pleasure and crying when annoyed. He could smile. There was a slight right abducent paresis and the face was asymmetrical because of weakness of the left face at rest and on movement. Swallowing was normal.

The child moved the right arm more than the left but neither as freely as a normal baby. The legs were little moved. The Moro response was present. When the child was held upright with the head flexed, the legs took up a position of slight flexion at the hips, extension at the knees and severe plantar flexion at the ankles. There was some scissoring in the thighs. The upper limbs tended to be flexed.

When the child was held with the head extended and the head was briskly extended, immediate hyperextension of the neck and back resulted. The limbs were also extended, the upper limbs adducted at the shoulders, extended at the elbows and flexed at the wrist and fingers. The forearms were pronated. The lower limbs showed full extension at the hips, the knees were extended and there was a position of equinus of the foot bilaterally. The child was extremely rigid in this position. By jerking him suddenly so that head extension resulted, the opisthotonic attitude described above was immediately produced.

There /

There was a suspicion of increase of tone in the left arm and a definite increase of tone in the legs bilaterally of rigid type. The tendon jerks were all slightly brisker than normal. Stretch responses could be elicited from both lower limbs and the left arm.

Examination at the age of 3 months showed some changes. The child was 24" in length, the head circumference was $14\frac{1}{2}$". There was a definite right abducent paresis and a slight left abducent paresis. There was a definite left sided facial paresis. The child could hear. Swallowing appeared to be normal.

The dystonic attacks could still be produced by extending the head rapidly. But they were less easily produced than on the previous examination. There was a generalised increase of tone in the lower limbs and in the left arm. The right arm showed a less marked increase of tone. The increase of tone was rigid in type. Stretch responses could be elicited in the left upper limbs and the lower limbs. The tendon jerks in these limbs were much exaggerated compared to those of the right upper limb. The Moro response was absent. The child showed a marked tendency to scissoring and extension of the lower limbs when held upright and the left upper limb showed a position of extension as an associated movement very readily. The right arm was the only limb which was moved at all freely.

Sensory findings could not be tested in great detail, but pin pricks appeared to be appreciated in all areas.

transition of three of these cases to the stage of rigidity within a period of months.

Two of the cases were triplegic, four were tetraplegic; all were considered to be severe or moderately severe cases of diplegia. All the children were under the age of four years and three of them were under the age of 12 months. None of them was able to sit during the stage of dystonia and manual manipulation was very infantile and clumsy.

One case was observed first in the stage of hypotonia. Dystonia commenced at the age of 11 months and was still present at the age of 2 years and three months. By this time some rigidity of the legs was evident. (Case 13).

Another child was seen at the age of six weeks about seven days after the first dystonic phenomena had been noted. These were not marked but occurred up to 12 times a day when he was handled. By the age of three months they had almost disappeared, and rigidity, which had not been present at the time of the first examination, could be appreciated in the adductors and hamstrings in the lower limbs. Obviously such a short stage of dystonia is likely to be missed when mothers are questioned years later. (Case 138).

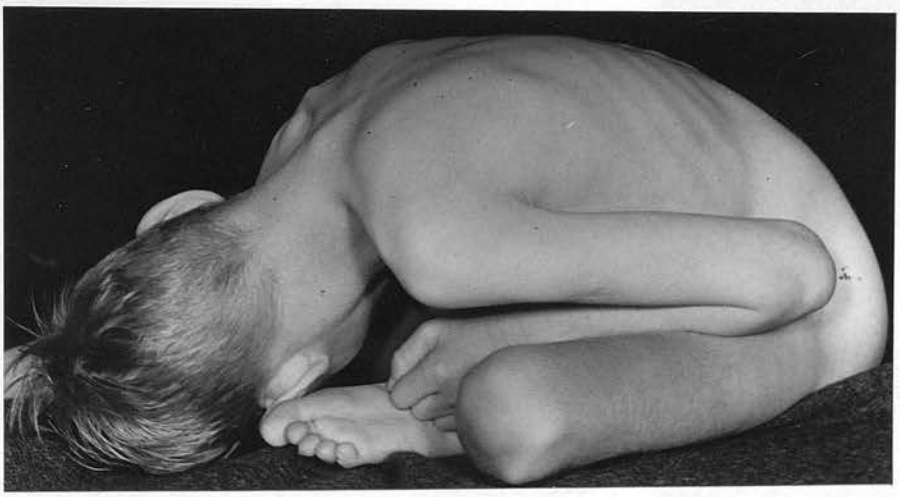
One case was seen in which dystonic phenomena persisted in spite of increasing motor skill. In the two previously quoted cases dystonia was a transient phenomenon, but in Case 56 the child was much more severely affected, and it seems likely that dystonic movement may persist.

Cases /

Figure 12.

Postures and reflexes in the dystonic stage of diplegia. With head flexed in the prone position there is generalised hypotonia.(a).The Moro reflex is positive at the age of eleven years.(b). Case 34.

a.



b.

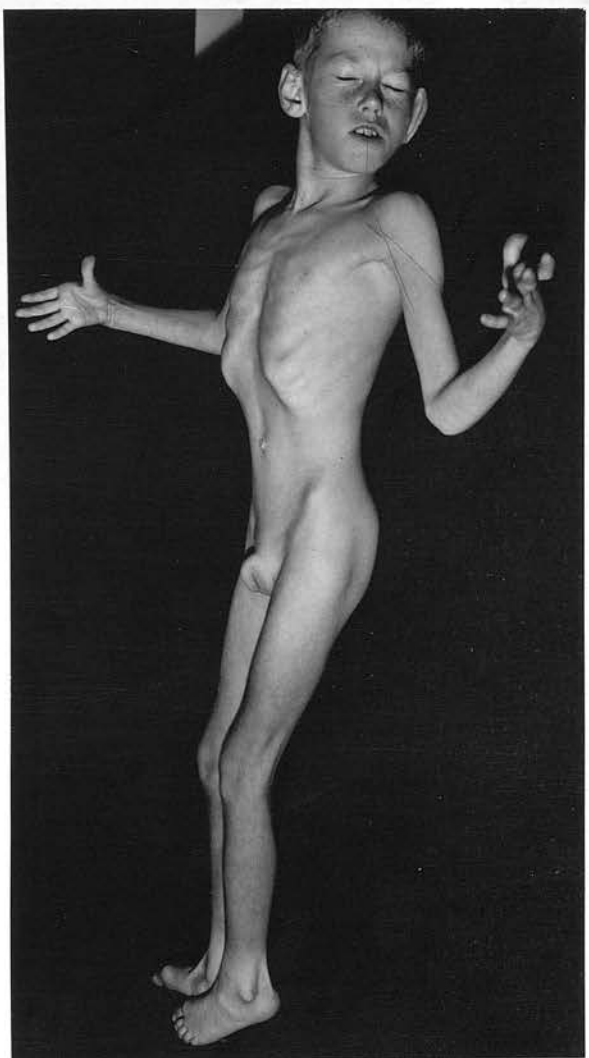
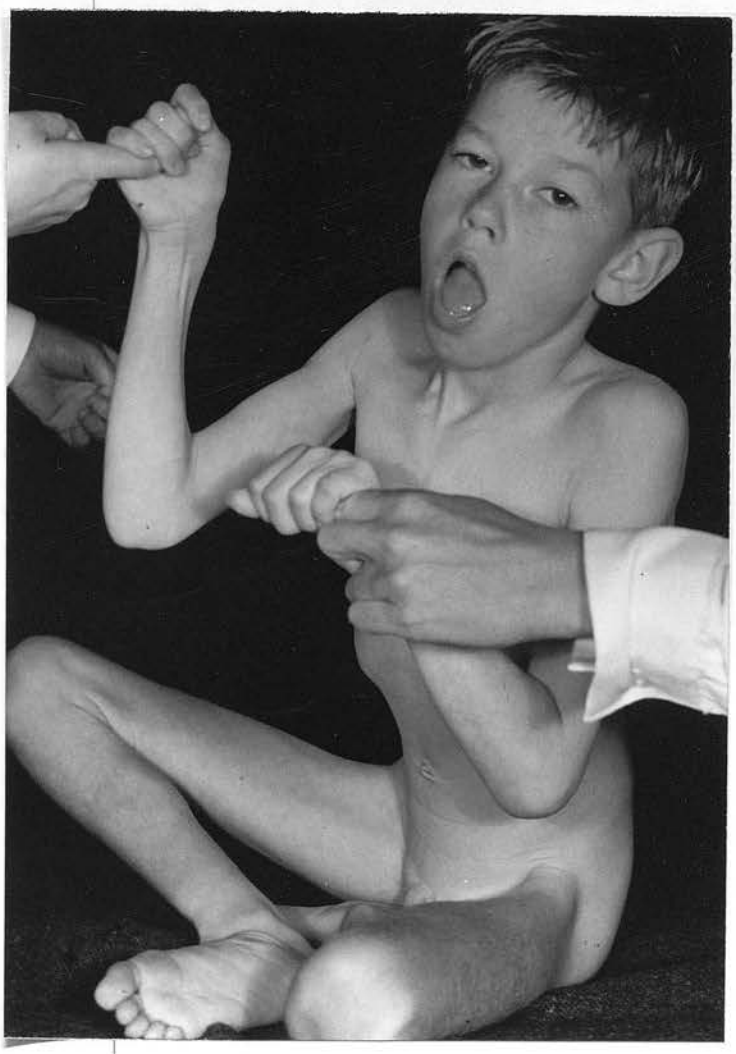
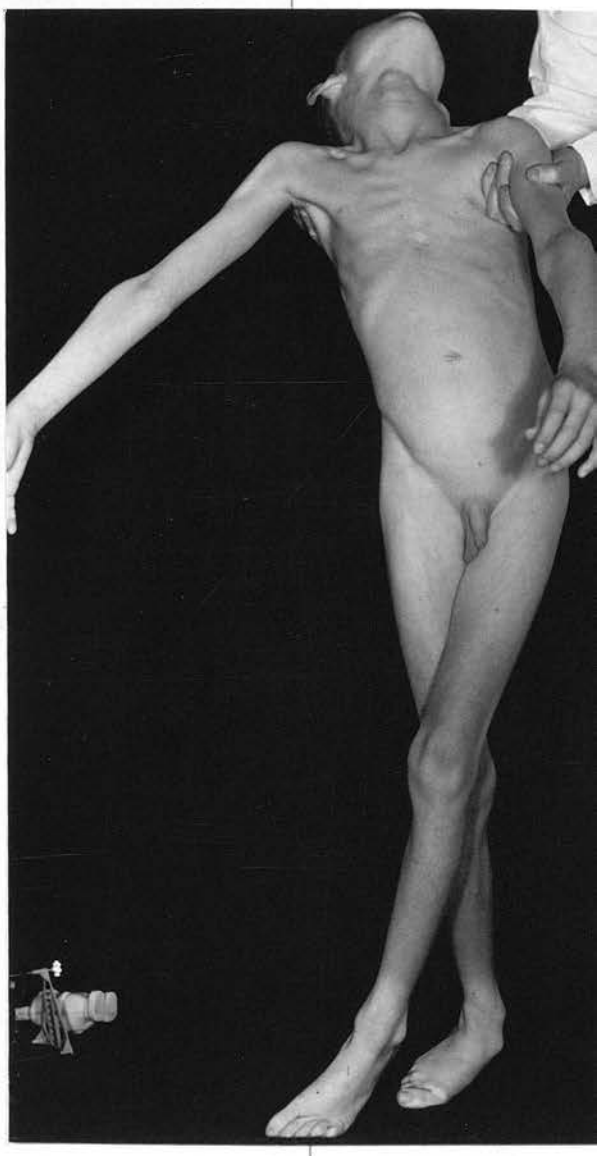


Figure 12 .(Continued).
Postures and reflexes in the dystonic stage of diplegia, (case 34) when the earliest signs of the rigid/spastic stage are making their appearance.Grasp reflexes,(a).The effect of extending the head with the child in the erect position and pressure on the soles of the feet.



a.



b.

TABLE 123

The ages at which 79 patients with diplegia walked without support

Distribution of paresis	Tetraplegia	Triplegia	Paraplegia	All cases
Age				
Under 18 months.	0	2	7	9
Over 18 months, under 3 years.	4	6	15	25
Over 3 years and under 6 years.	9	8	3	20
Over 6 years and under 8 years.	3	1	0	4
Over 8 years.	2	1	0	3
Not walking	5	3	3	11
Unknown	4	2	1	7
	27	23	29	79

Cases in the Rigid and Spastic Stage.

Of the 79 cases classified as diplegic, 72 were in the stage of rigidity or spasticity, with or without contracture, at the time of examination. The numbers of paraplegic, triplegic and tetraplegic cases and their classification into severe, moderately severe, and slight categories is shown in Table 122 . That the degree of functional impairment is roughly related to the number of limbs involved is shown in Table123,124 where the times at which the child first walked and talked is noted.

That the majority of cases of tetraplegia are unable to walk unsupported before the age of three years indicates how severely these children are handicapped.

The Impairment of Voluntary Movement.

The causes of the impairment of voluntary movement will be considered under the headings of loss of voluntary power, involuntary movements, rigidity, contracture and other disturbances. It is even more difficult in diplegia than in hemiplegia to assess to what degree the impairment, the power and co-ordination of voluntary movements is due to each of the various factors involved.

Loss of Voluntary Movement Patterns.

Loss of voluntary movement patterns occurs in even the mildest cases of diplegia and in mild triplegic and tetra-plegic cases it is the main cause of impairment of function in the upper limbs.

The /

TABLE 124

The age at which 79 patients of diplegia said their first words

Distribution of paresis	Tetraplegia	Triplegia	Paraplegia	All cases
Age				
Under 18 months.	3	9	18	30
Over 18 months, under 3 years.	9	7	7	23
Over 3 years, under 6 years.	3	4	3	10
Over 6 years, under 8 years.	2	0	0	2
Over 8 years.	0	0	0	0
Not walking.	8	1	0	9
Unknown	2	2	1	5
	27	23	29	79

The loss of voluntary movement patterns may be regarded as a result of retardation or failure of the normal development of voluntary motor control. Thus the motor control in cases in the hypotonic stage of diplegia resembles that of the new-born child. In the dystonic phase and the early rigid stage severely affected patients show a development of motor control similar to that found in the normal child up to the age of about 8 or 9 months. The hands are beginning to be utilised in conscious activities but thumb and forefinger movements are little used. The onset of the stage of spasticity coincides with the appearance of more highly developed adult types of movement patterns which are impaired by the motor disability.

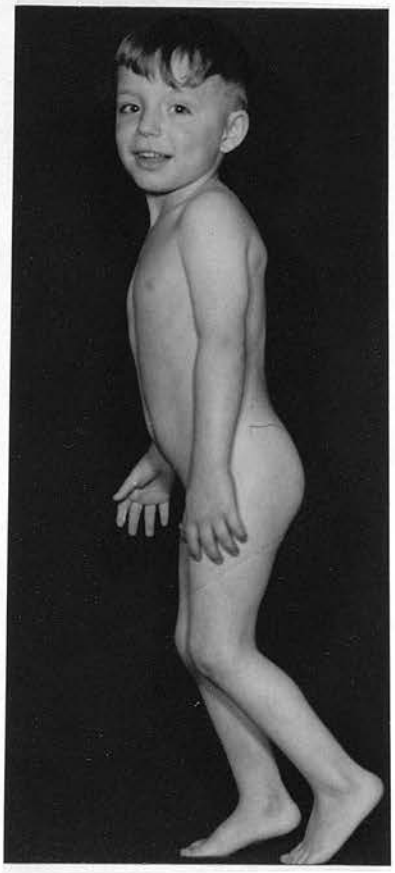
Impairment of voluntary movement patterns and power is most evident in later developed movements and in the periphery of the limbs. Movements of the thumb, extension of the fingers, wrist, and supination of the forearm are movements that are always weakened and sometimes limited in triplegia and tetraplegia. Voluntary toe movements are almost always absent or grossly impaired in diplegia. In early cases without considerable contracture it may be possible to demonstrate that dorsiflexion of the foot is limited in extent though no local cause for the limitation is apparent. There is an apparent loss of the movement pattern.

In severe cases of tetraplegic paresis, the impairment of voluntary power is evident, even in movements of quite primitive nature and of the proximal parts of the limbs. Unfortunately from the clinical point of view the loss, or

more /

Figure 14.

The gait in rigid/spastic diplegia. This relatively lightly handicapped paraplegic patient walks with feet in equinus and shows the typical leaning forwards attitude. (He also shows charcteristic internal squint.)



more accurately failure, to develop of actual movement patterns is overshadowed by the impairment of movement which results from later developing rigidity, involuntary movements, rigidity and contracture.

The loss of movement patterns is seen more clearly in cases of diplegia with cerebellar ataxia where rigidity tends to be less marked and in which very gross loss of the ability to move the fingers, toes, hands and feet voluntarily may be observed.

Associated Movements.

The impairment of motor control that results from associated movements is due to three types of movement in diplegia. Movement of the dystonic type, and athetoid movements similar to those found in hemiplegic cases, are common. Movements of dystonic type as a cause of impairment of motor function are, of course, most marked in the dystonic stage of diplegia and have been discussed in the section dealing with this phase of the development of the condition.

Sometimes, however, involuntary movements recognisably of similar pattern to the more extensive dystonic movement of the dystonic stage, persist into the rigid phase and are severe enough to cause severe impairment of voluntary movements. The stimuli of the erect position, pressure on the soles of the feet and the mixture of postural control and voluntary movement found when severely affected children began to walk were sufficient to elicit these involuntary movements in a number of cases.

In /

In these cases there was a very marked tendency for sudden extension of the neck, back and limbs to occur whenever attempts were made to take a step unsupported. Balance was immediately lost and retardation in achieving unsupported walking balance was often severe, though when the child was lifted so that the stimulus to involuntary postural movements of pressure on the soles of the feet was absent, walking movements of the legs might be perfect. (Case 34).

Only when the degree of motor control was sufficient to overcome the tendency to neck extension caused by pressure on the soles of the feet, the assumption of the erect position and massive walking movement was unsupported walking achieved. The involuntary movements of extension tended to be especially severe in all cases in which all four limbs were markedly involved, and in these children the use of an upper limb might be grossly restricted because of them. Objects could be grasped only very clumsily and sometimes the extension of the arm was severe and sudden enough to result in it being thrown out of the hand once grasped.

It was interesting to observe that on examination it was possible to produce isolated movements of extension in the limbs of the type described in four of the cases classed as paraplegic, six of those classified as triplegic and nine of those classified as tetraplegic, and this in spite of the presence of contracture in the latter cases. The most potent stimuli were found to be extending the head and neck, exerting pressure on the soles of the feet and maintaining the child in the /

Case 34.

A case of severe tetraplegic rigidity with marked involuntary extensor movements when feet were on the ground.

Male, born 1949, the fourth child of a mother aged 36 at the time of delivery. The first child was a forceps delivery, the second born after a labour of less than one hour. Both were born at term. Both healthy. Third child stillborn as a result of concealed/revealed haemorrhage from placenta praevia.

The mother was well in her fourth pregnancy. She was given ergot tablets at term and an injection of pituitrin. One hour after the latter the child was born precipitously while the mother was sitting in a chair, as the result of a single pain of great severity. The child weighed 7 lb.

The child did not breathe for about 20 minutes and did not cry for over one hour and then very feebly. He was blue for almost two days. He had to be tube fed for over one week. Thereafter he was bottle fed but swallowing was very difficult and he often coughed up milk during feeds. At the age of 3 or 4 days he began to show sudden attacks of stiffness in which he arched the back and neck and extended the limbs. "Like a stiff doll, not like a baby at all". The attacks occurred whenever the child was moved, when he choked, and sometimes apparently when he was in his cot alone. The attacks continued until the age of two, by which time he was able to hold up the head. He succeeded in sitting with support at the age of $3\frac{1}{2}$. At the age of 4 he began to hold toys in the left hand but movements of this were very clumsy and he frequently dropped them.

His limbs were noted to be becoming stiff shortly after the age of one year. The stiffness appeared first in the legs and has remained more marked in them than in the upper limbs, though these are also affected. The stiffness is now marked and at times he is "like a matchstick man with his arms screwed on the wrong way round, and his hands pointing to the back".

On examination at the age of 4 he was $33\frac{1}{2}$" high and head circumference was 18". His teeth were very decayed. He could obey simple commands; trying unsuccessfully to speak; drooling and swallowing infrequently. Appeared to see and hear normally. Bilateral abducent paresis, more marked on the left than the right. The face was rather expressionless and immobile, but not asymmetrical. Swallowing was slow, infrequent and obviously difficult. Tongue movements in all directions were slow and restricted in range. The grasp of the hands was fair. The whole hands were used and there was no attempt to hold objects with the finger and thumb alone. The thumbs were contracted into the palms though not severely. The /

The forearms showed bilateral pronation contracture. Dorsiflexion of the feet was impossible on account of contracture of the tendon Achilles.

There was generalised rigidity of the limbs, much more marked in the lower limbs than the upper. The tendon jerks were very greatly increased in all the limbs. The knee and ankle jerks were more increased, relatively, than were those of the upper limbs. The plantars were bilaterally extensor. Stretch responses were evident in all limbs.

Whenever he attempted voluntary movement of the fingers severe athetosis of the fingers and wrists was evident bilaterally. Whenever movements of the whole upper limb were attempted there was a marked tendency to associated extensor movements. These were severe in type and resulted in the upper limbs being adducted at the shoulders, extended at the elbows and pronated at the forearms. The wrist and fingers were extended ("arms on the wrong way round"). The legs were extended at the hips, knees and the feet were plantar flexed. Slight scissoring was apparent.

When he was placed on his feet in the upright position, the head was suddenly extended, the back arched, the limbs became markedly extended. All attempts at standing balanced were vitiated by a form of dystonic attack. When he was held off the ground, however, the dystonic position was not in evidence. The legs were placed in a similar position to that found when the feet were placed to the ground. But no extension of the back or neck was evident. The upper limbs were not strongly extended but rather tended to be held in flexed positions.

It was impossible to produce dystonic attacks by extension of the head unless the pressure was exerted on the soles of the feet, either by placing a hand against them, or by placing them on the ground.

Sensory findings could not be tested fully but pin prick appeared to be appreciated in all areas.

Case 147.

A case of moderately severe triplegic paresis with athetoid movements in the upper limbs.

Male, born 1950, the first child of healthy parents. Mother aged 31 at the time of delivery. She had severe pre-eclampsia in the last six weeks of her pregnancy. She went into labour at term and was admitted to a Nursing Home, but after a few hours pains ceased and she was given pituitrin injections to restart them. These injections, however, were unsuccessful, and she had an artificial rupture of membranes three days later, after which labour pains commenced. The child was delivered by mid-cavity forceps delivery.

Birth weight was 6 lb. 12 oz. He was born crying and seemed well until the seventh day when he had a generalised convulsion. Twitching of the limbs continued for four days. Thereafter he seemed to improve and was discharged from the Nursing Home at the age of 4 weeks. Apart from crying a great deal he appeared to be normal. At the age of six weeks, however, he sometimes seemed stiff in the mother's arms. This was most evident when he was being bathed and the head was held back. Mother does not remember position of the limbs.

At the age of three months he was able to hold up the head. By this time the legs were definitely stiff and the right hand seemed to be stiff, the fingers clenched and the limbs moved less freely than the left. The child could sit without support at the age of 15 months and shortly after began to propel himself along with the left arm, while in the sitting position. He was able to walk with support at the age of three years. He had begun to speak at the age of 9 or 10 months.

On examination at the age of $3\frac{1}{2}$ he was an intelligent spoilt boy with spastic triplegia. Height was $34\frac{1}{2}$". Head circumference was 20".

His speech was normal except for slight slurring of consonants. Its content was good. There was bilateral abducent weakness. Slight right facial weakness was evident on movement only. The tongue deviated to the right on central protrusion.

There was impairment of power in all the limbs. This was moderately severe in the legs and right arm, but much less severe in the left upper limb. Full abduction of the thumb and supination of the forearm was limited on the right and the latter slightly limited on the left. Dorsiflexion of the feet was rather severely limited bilaterally. Movements of the left hand were rather clumsy but probably not outwith the range of normality. In the left hand, however, all /

all movements were accompanied by a marked degree of athetosis of the fingers which were severely hyperextended. The wrist showed some involuntary extensor movement at the same time as the fingers. The involuntary movements of the fingers and wrist were slow and writhing in type. He was able to grasp with the left hand when made to, but was unable to release the grasp.

There was a marked spastic increase of tone in the left arm and in the lower limbs, the increase in the latter being more severe. There was a slight spastic increase of tone in the right arm. The biceps, triceps and supinator jerks were much increased in the left arm and slightly in the right. The knee and ankle jerks were much increased bilaterally. The plantar responses were extensor.

Sensory findings could not be tested fully, but no impairment was demonstrated.

the erect position. In all these cases and in all the tetraplegic cases the tonic neck reflex could be relied upon to produce a more or less easily elicited extension in the limbs on the side from which the head was turned.

In a few cases the movements of dystonic type occurred until very late in the rigid and spastic stages of diplegia, and resulted in great functional impairment of voluntary movements.

In most cases, however, the dystonic types of involuntary movement disappeared when spasticity of the limbs became apparent, after a longer or shorter period of time. In many cases, however, they were superseded by athetosis in the spastic limbs leading to almost equally severe derangement of function. This athetosis is almost identical to that described in hemiplegic patients. In some tetraplegic children it is bilateral; in others one limb may be spared or show the more primitive type of extensor movement. As in hemiplegia the degree of functional impairment it causes varies greatly. (Case 147).

The choreoid type of associated movement which occurs occasionally in hemiplegia also occurs, but much less frequently in patients suffering from diplegia in the spastic stage.

Rigidity of the limbs.

A cause of limitation of voluntary movement not found in cases of hemiplegia is generalised rigid increase of tone not due to contracture. This is almost invariable in cases of diplegia /

diplegia of moderately severe or severe degree. Indeed the earlier authors doubted if there was in fact any real loss of movement patterns in diplegia, and attributed the limitation of voluntary movement only to rigidity (Little, 1862; Freud, 1893).

The fact remains that rigidity is a very important cause of loss of range and power of voluntary movement in paraplegic, triplegic and tetraplegic cases. The rigidity of the limbs appears before the dystonic attacks have ceased, or in paraplegic cases with normal milestones about the age of 3 - 4 months. It is usually first evident in the thighs and knees, but rapidly spreads and increases in the lower limbs, and involves the upper limbs also. Initially marked variations from day to day and minute to minute may be found. When the child is excited, especially when feeding, the rigidity may be very pronounced. When the child is held in positions which are liable to produce dystonia in susceptible children, the rigidity is accentuated. The most potent means of accentuating rigidity has been found to be to extend the head. The easiest way to produce diminution in rigidity has been found to be to flex the head strongly. This latter manoeuvre has been much utilised by some schools for producing muscular relaxation in diplegic children (Bobath and Bobath, 1950).

In the same way as the assumption of the upright position, pressure on the soles of the feet and attempting voluntary movement produces associated extension positions in the limbs so they also tend to produce generalised increase of tone in the muscles of the affected limbs, whether or not, in fact, dystonia /

dystonia has been a feature in the individual patient.

The rigidity is always much more pronounced in the lower limbs than the upper, whether the case is slightly or severely affected. In moderately severe and severe cases it may be enough to inhibit all attempts voluntarily to move the limb.

The reflex nature of the rigidity is shown by the fact that voluntary movements may be rendered quite impotent by its presence in some positions of the child, whereas movements may be quite full in range if the child is placed in positions where his rigidity is diminished. The following case had the greatest difficulty in walking at all because of the extremely severe rigidity of his legs. Yet when placed on his back with the head strongly flexed the range of leg movements was good and their power only moderately impaired. The rigidity had been much reduced by the manoeuvres employed. (Case 25).

<u>The Spastic Phase of Diplegia and the Importance of Contracture in Limiting Voluntary Movements</u>.

The changes in clinical findings which occur with the onset of spasticity in cases of diplegia are fascinating. The stage of the disorder at which spasticity becomes evident usually coincides with the child's first attempts to use the thumb and forefinger, and with his ability to control the postures of his limbs so as to allow more efficient handling of objects to be possible.

At the same time voluntary movements cease to result in the associated movements of pure extension. Some flexion is first evident in the elbows and over a period of weeks or months /

Case 25.

Severe spastic tetraplegia illustrating varying degree of rigidity in different postures.

Male child, born in 19 , the first of four children, born to a mother aged 19 at the time of delivery and her healthy husband. Three subsequent children born after normal pregnancy and delivery are well. Mother and maternal aunts and grandmother are left handed. Mother was well during pregnancy until four weeks before the expected date of delivery when she had a severe vaginal haemorrhage. She was admitted to hospital where a placenta praevia was found and Willetts forceps were applied to the foetal head. Delivery occurred after a labour of two hours and was accomplished under anaesthesia. The placenta showed numerous infarcts and weighed 1 lb. 5 oz. The child was limp following delivery but responded to oxygen, though on admission to the premature nursery was said to be "rather murmury". The birth weight was 5 lb. 15 oz. After two days in the premature nursery the child appeared to be normal and breast feeding was begun. He was discharged on the thirteenth day.

He was said to be a good baby. It was noted that the left eye was turned in. He sat with support at the age of 8 months and alone shortly afterwards. He began to crawl at the age of two years, and when he did so it was found that he pulled himself along with the arms, pulling his legs behind him stiffly "like a tail". He was able to stand at the age of $3\frac{1}{2}$ and to walk without support at the age of $4\frac{1}{2}$. When he began to walk he was very unsteady and fell a great deal because he kept putting one foot in front of the other and tripping. Gradually his gait improved and by the age of 5 years he was able to walk without falling more than occasionally, though he still proceeded on the toes, not putting the heels to the ground. With lengthening of the tendon Achilles operatively at the age of $5\frac{1}{2}$ the gait improved further and he managed to get the heels to the ground.

Mother thought that he was rather slow to use the hands and used the right hand before the left. He always handled objects poorly, and at the age of 11 could still not do all his buttons or his laces. His right hand was better than his left. He said his first words at the age of 18 months and his speech developed normally thereafter. At the age of 6 he went to a school for the physically handicapped and has done moderately well there, learning to read and write without apparent difficulty. I.Q. 82.

Of quiet, placid disposition, but able to play successfully with children of his own age.

Examination. /

Examination. Height 55½". Head circumference 21½". He was slow in response to commands and in answer to questions but was accurate. His comprehension was good but his speech, though of fair content, was rather slurred and jerkily pronounced. There was a slight right facial weakness shown by lag on voluntary and emotional movement, though the face was symmetrical at rest.

The power and co-ordination of the limbs was impaired. The impairment was more marked in the legs than the arms. There was limitation of passive supination of the left forearm, but other passive and voluntary movements were full though supination of the forearm, extension of the fingers, wrists and elbows, and abduction of the shoulders were weak bilaterally, though more impaired on the left than the right.

Fine and rapid finger movements were poor bilaterally, and though he could handle small objects he used the whole hand in an infantile way without especial use of the thumbs and forefingers. The finger movements were better controlled on the right than the left.

There were flexor contractures of the hips, knees and ankles preventing full hip and knee extension and full dorsiflexion at the ankles. The contractures were more severe on the left than the right. The gait was spastic with scissoring of the legs, flexion and adduction of the hips, flexion of the knees and a tendency to equinus position of the feet. As he walked he leant forwards and the arms were in perpetual motion, tending to be held adducted at the shoulder, flexed at the wrists and fingers, pronated at the forearms, and flexed to variable degrees at the elbows. He had great difficulty in maintaining his balance when he initiated his first step.

There was a marked increase of tone in the limbs. This was moderate in the right arm, rather more severe in the left arm and very severe in both legs. The tone was spastic with underlying rigidity. Stretch responses were present in all limbs. The biceps, triceps and supinator jerks were moderately increased, more on the left than the right, and the knee and ankle jerks were much increased with clonus at both knees and both ankles. The Hoffmann signs were present bilaterally. The plantar responses were extensor on right and left. The fingers showed some hyperextensibility bilaterally.

The tonic neck reflexes were well marked. When he used either upper limb forcefully the contralateral arm also showed movement and tended to assume a position which it would have shown in generalised opisthotonus. When the head and trunk were suddenly and forcefully hyperextended a generalised position of opisthotonus was momentarily assumed. When he was /

was laid supine and his head flexed forwards firmly there was a remarkable increase in the freedom of voluntary movement, and on examination the rigidity of the limbs was found to be much decreased. Apart from some restriction of the ability to dorsiflex the feet and only minimal abduction at the hips, the range of movement in the lower limbs was full and surprisingly powerful. In the upper limbs only minimal supination was possible, but other movements were full in range and rapidity of finger movements was noticeably increased. No abnormal sensory findings were elicited.

months the flexion spreads and becomes more marked so that the whole of the involved upper limbs assume positions similar to those found in hemiplegic patients on voluntary movements.

It is difficult to see that the appearance of spasticity itself has any effect on the functional use of the involved upper limbs. The lack of power and range of movement is evident before spasticity appears. In the lower limbs the greater difficulty in moving from the hips does seem to have some deleterious effect in some cases, and in two the histories suggest that children beginning to walk temporarily retrogressed at this stage.

The great importance of the appearance of spasticity from the functional point of view is the rapidity with which it may be complicated by severe contracture in the limbs. The same factors as favour the appearance of spasticity in hemiplegia also favour it in cases of diplegia. Since the history in diplegic cases tends to be less dramatic than in many cases of hemiplegia, these cases tend to remain without adequate medical treatment for longer. In fact, though the majority of cases of diplegia were taken to their doctors very shortly after the appearance of their first symptoms, there was an average delay in their receiving adequate medical treatment of almost two years. In almost all cases contractures of some severity were present by this time.

As in hemiplegia, the contractures tend to be most marked in the periphery of the limbs than the proximal. This leads to a very interesting phenomenon in diplegia of hands, clumsy, but /

but otherwise apparently unaffected becoming relatively useless in quite a short space of time. Whereas before the onset of contracture the child usually finds abduction and extension of the thumb and fingers difficult, when contracture ensues, these movements may become impossible in a matter of a few weeks.

Pronation contracture of the forearms in those with upper limb involvement is invariable in those with much spasticity, the elbows and shoulders are less affected. In contrast to hemiplegia the legs are more involved than the arms. The feet become fixed in a position of plantar flexion, the legs and hips in flexion, or variable degrees of severity.

The age of the appearance of contractures is extremely variable. As has been noted, the spastic phase may be delayed in its appearance for up to four years, and even longer. Some contracture has been noted as early as six months, though to find it at this early age is unusual. Very severe contractures may be present by the age of two years.

From the point of view of function, however, the most severe results of contracture are often found in the hands, where movements already weak and poorly co-ordinate are further grossly impaired.

Of the 79 cases of diplegia in the series, 28 had already had tendon lengthening operations by the time of examination, others were obviously going to require them.

Yet /

Yet it was apparent that in other patients the necessity for operation had been avoided by early diagnosis and physiotherapy though their paresis was as severe as in the former group.

In the legs it was often extremely difficult to assess to what degree the stiffness of the legs was due to true muscular rigidity of reflex origin and how much to contracture. This was especially true of the marked plantar flexion of the feet that was found in severe cases of diplegia. The degree of flexion contracture evident at the hips and knees was not a reliable guide as it depended so much on the degree of disuse to which the limbs had been subjected.

As was to be expected, contracture was more marked in older patients than in younger. As in hemiplegia, contracture appeared to be somewhat exacerbated about the time of puberty (Féré, 1896).

Dwarfing in Diplegia.

The marked dwarfing of the pelvis and lower limbs evident in diplegic patients was noted by the early observers of the condition (Little, 1843; 1862; Gowers, 1888; Freud, 1893). Its severity roughly parallels the severity of the paresis of the lower limbs in the majority of patients.

In view of the fact that the most severely affected limbs show marked retardation of growth in diplegia, it might be expected that in severe cases of triplegia dwarfing of the affected upper limb as well as of the lower would be apparent.

In /

Figure 15.

Illustrating the marked dwarfing of the pelvis and lower limbs characteristic in diplegia.

In fact this was not found. Only one case of triplegia showed apparent shortening of more than half an inch in the more affected upper limbs compared to the less affected. This is in accordance with the observations of Freud (1897).

Why dwarfing of the affected upper limb should occur in hemiplegia and not in triplegia is difficult to see. It is the more difficult to account for it when the similarity of some of the other findings in hemiplegia and triplegia are considered. Many of the latter, in fact, have a superficial appearance of paraplegia with superimposed hemiplegia. Spasticity of the upper limb rather than rigidity is evident; the tendon jerks are much increased; the Hoffmann response is frequently psoitive; athetosis may be present; the associated movements are flexor in type; the contracture is similar to that found in hemiplegia. Yet dwarfing in the more affected upper limb is exceptional in triplegia and usual in hemiplegia.

It is interesting also to note that in triplegic patients it is unusual for there to be significant temperature difference between the hands.

In the majority of cases of diplegia the head was smaller than was to be expected on the basis of current tables of head size. Since over one third of the patients were prematurely born, however, more detailed study of the results of head measurements must await the availability of comparable Edinburgh figures. Seven cases, four tetraplegic, two triplegic and one paraplegic, showed a difference of

half /

half an inch or more in the cranial hemicircumference. A much larger number showed quite marked plagiocephaly, which most disappointingly, gave very similar measurements for each hemicircumference.

Other Neurological Findings in the Limbs in Diplegia.

Vaso-motor changes in the limbs in diplegia were sometimes evident in the feet but rarely in the hands and in no case was marked difference between the sides noted. In most cases the feet felt colder than the hands, and pallor was sometimes evident but the findings were not striking.

In one case only was definite sensory imapirment demonstrated, a loss of joint sense in the lower limbs. Very slight loss of joint sense was almost universal in those whose limbs were immobile but in no case receiving routine physiotherapy was even this discovered.

Neglect of the more severely affected limb, similar to that found in hemiplegia was not found to be a feature of diplegia though in a few cases detailed questioning of the mother gave the impression that neglect of the more severely affected side might be present transiently following a fit, expecially in triplegic patients.

Mental Impairment in Diplegia.

As in hemiplegia, the estimation of intelligence by tests in diplegic children is fraught with uncertainties. The success of the child in achieving social adaptation and educational standards is probably equally fallible as a guide

to /

to intellectual impairment (Dunsdon, 1952).

The schooling received by diplegic children and their intelligence as estimated by different psychologists and tests in various circumstances are shown in Tables 125, 126 . It must be emphasised that the test results are not to be taken as accurate representations of the actual intelligence of all cases because the method of testing varied so greatly. Moreover 20 patients, four of whom were in normal schools, were not tested for various reasons. The most that can be taken from the results is that the numbers of children with severely impaired intelligence is greater in tetraplegic cases than in paraplegic, and that a higher proportion of the more severely paretic cases are ineducable, than of the less severely affected. The small number of children with diplegia attending normal schools is striking.

The severity of intellectual impairment as measured by intelligence testing was not always a true reflection of a child's inability to achieve some success in the social sphere. The achievement of some institutional cases was surprising in view of the low measurements of intelligence obtained on testing.

The remarks made about the value of milestones in assessing the degree of mental impairment held true for diplegia. The most reliable single criterion of mental impairment judged from milestones was delay in talking, and in a number of children with quite severe speech retardation only slight impairment of intelligence was found. Thus even speech is not /

TABLE 125

The intelligence quotient in 79 children with diplegia

	No. of cases	115+	100-114	85-99	70-84	55-69	Under 55	Untested
Paraplegia	29	1	5	4	6	7	1	5
Triplegia	23	2	2	0	5	3	3	8
Tetraplegia	27	0	0	2	4	3	11	7
All cases	79	3	7	6	15	13	15	20

TABLE 126

Schooling in 79 patients with diplegia in childhood.

	No. of cases	Normal Schools	Schools for the physically handicapped	Schools for the mentally handicapped	Ineducable	Not yet at school
Paraplegia	29	7	9	5	0	8
Triplegia	23	3	3	5	4	8
Tetraplegia	27	1	0	6	9	11
All cases	79	11	12	16	13	27

not a constantly reliable measure.

Aphasia.

Aphasia was recognised in only four cases of diplegia. Three of these were triplegic, two with predominant upper limb involvement on the right and one on the left. One case was tetraplegic. In no case was the aphasia severe.

A much larger number of mental defective cases of diplegia appeared to have either greater difficulty in comprehension than expression or vice versa. It is probable that a number of these cases did in fact have disturbances similar in type to those classified as true aphasic disorders, but that they were much slighter in degree.

Speech Defects.

Thirty-five children with diplegia showed speech defects of various types. The numbers occurring in paraplegia, triplegia and tetraplegia are shown in Table 127 . It will be seen that speech defects were more frequent in tri- or tetraplegic patients than in those with paraplegia.

For the purposes of classification stammer and hesitation were grouped together. They were infrequent. Retarded development of speech, most often manifest as inability to pronounce consonants, was relatively common. True dysarthria with characteristic patterns of defective pronunciation was practically confined to tetraplegic patients.

The severity of the speech disorder was assessed purely on the basis of the intelligibility of the child's speech to his /

TABLE 127

Speech defects in 79 patients with diplegia

Articulatory defects

	No. of cases	Stutter or hesitation	Articulatory Defects: Mild	Moderately severe	Severe	No more than one or two words.	Total speech defects.
Paraplegia	29	0	3	1	2	1	7
Triplegia	23	4	4	3	4	1	14
Tetraplegia	29	0	2	4	5	3	14
Diplegia	79	4	9	8	11	5	35

TABLE 128

Impairment of vision in 79 cases of diplegia

	No. of cases	Retrolental fibroplasia	Optic atrophy	Cortical blindness	Total
Paraplegia	29	1	1	0	2
Triplegia	23	3	0	0	3
Tetraplegia	27	2	2	4	8
Total diplegic patients	79	6	3	4	13

his family and to strangers. Mild speech defects were those which, though they sometimes made speech difficult to understand for strangers, did not make it unintelligible. Moderately severe speech defects were those which made what the child said more or less unintelligible to people outside the family. Patients with severe speech defects were unintelligible even to their parents and siblings.

Five patients, all severely affected, had such defective speech as to be unintelligible even when saying single words. Another eleven could not express themselves intelligibly when trying to form phrases. They are grouped as having severe speech defects in Table 127 . In eight patients speech defects were moderately severe, and slight in nine.

Cranial Nerve Involvement.

The proportion of diplegic patients showing cranial nerve abnormalities, with the exception of strabismus, is stated to be low in most surveys (Osler, 1889; Lovett, 1888). But some early work tended to confirm the present findings that cranial nerve abnormalities are not, in fact, common (Koenig, 1896).

Visual Defects. Complete or partial blindness was ascertained in 13 cases. In 6 cases retrolental fibroplasia was present. In three optic atrophy was present, and in four field defects were evident, presumably of cortical origin (Table 128) (Ingram and Kerr, 1954).

All the cases of retrolental fibroplasia were cases of severe /

severe paresis, one paraplegic, three triplegic and two tetraplegic in extent. All the children were born prematurely and all had a history of neonatal anoxia and oxygen therapy.

Optic atrophy with typical retinal appearances was observed in three cases, one paraplegic and two tetraplegic in distribution. Two of the three cases were blind except for appreciation of light. One had a degree of useful vision, but was severely mentally defective. Three had some useful vision.

Field defects were present in 4 cases. One case showed a complete homonymous hemianopia and had useful vision. The other three cases had vision in only one quadrant and the degree of vision, or the child's utilisation of it, was so poor as to be useless.

Of the 13 cases with blindness, partial or complete, five cases were considered to show a degree of useful vision, and eight to have no useful vision.

One case of tetraplegia showed the typical retinal changes of toxoplasmosis, but visual acuity and fields were normal at the time of examination.

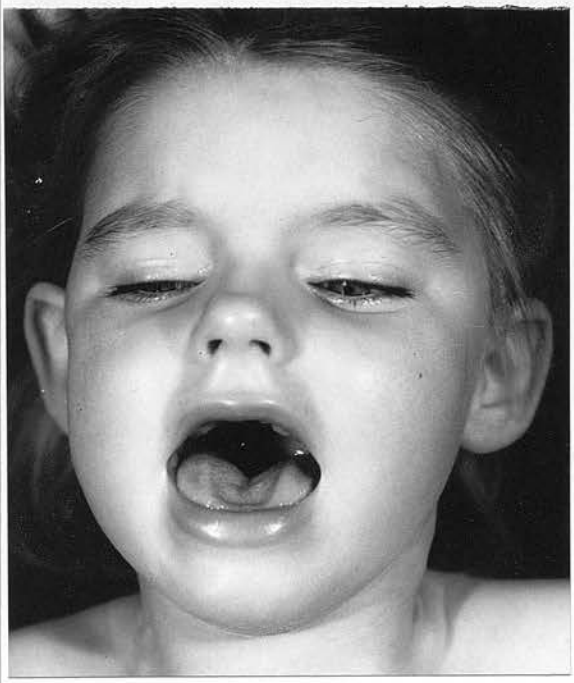
A number of cases showed refractive errors of various degrees of severity. Three cases were severely myopic.

Strabismus. The figures in Table 129 indicate a frequency of strabismus in diplegia in agreement with that noted by early authors. (Koènig, 1896; Freud, 1893). Thirty-four patients showed strabismus. The commonest type noted was convergent strabismus which was very severe in some cases and /

Figure 16 .

Somewhat atypical severe rigid/spastic diplegia with more bulbar involvement than usual.(a).Unsupported sitting,(b). Standing posture intermediate between rigid/extensor and spastic/flexor types.(c).Tonic neck reflex.(d).

a.



c.

d.

and slight in others. Strabismus was commoner in tetraplegic paresis than in triplegic or paraplegic paresis. It was commoner in those born prematurely than in those born at term. Since the strabismus tended to improve as the child grew older, in most cases it was more frequent in its occurrence and more severe in young patients.

In three cases there was bilateral abducent paresis of severe degree which appeared to be of central origin. The eyes could be made to abduct fully involuntarily, but only very slightly on voluntary eye movement.

In another four cases of severe symmetrical bilateral abducent paresis supranuclear paresis was suspected, but because of poor co-operation of the young children could not be confirmed on full examination. It is possible that supranuclear paresis is commoner in diplegia than is generally thought, but the difficulties of confirming its presence in young children are considerable. One case of supranuclear third nerve paresis with inability to close the eyes voluntarily or adduct the eyes voluntarily has been observed, but the child was not resident in Edinburgh and is therefore not included in the survey. The patient was a girl with severe tetraplegia. One patient with bilateral third nerve paresis in the survey was ssuspected of having a degree of supranuclear paresis. His eye movements were conjugate on involuntary movement, but not conjugate on voluntary movement and voluntary eye closure was very weak. He was a case of severe tetraplegia.

All /

All the cases with retrolental fibroplasia had unilateral abducent weakness. Strabismus was evident in all but one of those with partial or complete blindness.

Facial Paresis. In contrast to some of the earlier statements that facial paresis was unusual in diplegia, facial paresis was found in quite a high proportion of cases of diplegia. This is in agreement with the observations of Koenig (1896) and Freud (1893, 1897).

In the series thirty patients were found to show unilateral facial paresis and three, all cases of tetraplegia, bilateral facial paresis. In only five cases was facial asymmetry present at rest. In the remaining cases of unilateral paresis the face became asymmetrical only on movement. As with facial paresis in hemiplegia, a number of these patients showed overaction of the affected side on movements of expression, and underaction of the face on voluntary movements.

In the ten triplegic cases and 13 tetraplegic cases who showed unilateral facial paresis this was on the side of the most affected arm in all but one case. In those showing bilateral facial paresis it was usual for the paresis to be asymmetrical. In these cases the face tended to be somewhat mask-like and moved little on either voluntary or emotional movement.

Swallowing. A history of drooling and apparent difficulty in swallowing during infancy was elicited in many of the patients with tetraplegia and triplegia.

It /

It was difficult to distinguish between those cases in which the difficulty of swallowing was due to feeding mismanagement and which to organic disorder. But in nine, organic difficulty was demonstrated. Drooling was a prominent feature. Seven of the nine cases who showed difficulty in swalling were tetraplegic, seven were over the age of three, and five were severely mentally defective. All but one showed concomitant facial paresis, either unilateral or bilateral, and three showed evidence of upper neurone paresis of the tongue. All those with difficulty in swallowing had speech defects.

In some cases the difficulty in swallowing was so severe that weaning had proved impossible for considerable periods. In some cases swallowing was only evident once in three minutes or more.

The Tongue. The tongue showed defective movement in seven cases, three with triplegia and four with tetraplegia. In three cases the movements appeared to be limited and weak in all directions. In four cases the weakness was asymmetrical. Three of the cases showed difficulty in swallowing and all had facial paresis. All but one of the cases with involvement of the tongue showed defective speech.

Other Cranial Nerve Disturbances. One case showed impaired ability to elevate one shoulder. She was a case of paraplegia with mental defect, epilepsy and a family history of /

of neurofibromatosis.

Epilepsy.

Epilepsy is a frequent manifestation of diplegia. Many different forms of epilepsy occur and the form of the attacks differs greatly in different cases.

The types which occurred and the frequency of their occurrence in the different forms of diplegia are shown in Table 130 . It will be observed that 21 cases had a history of epileptic attacks. Twelve of these had grand mal only, 6 had grand mal and petit mal, two had Jacksonian attacks, and three had myoclonic jerks only. In two of the latter grand mal also occurred.

Grand mal alone or in combination with other forms of epilepsy was present in 20 cases. The time of the occurrence of the first fit, the time of the occurrence of the last fit and the frequency of attacks are shown in Table 131 and 132 . It will be seen that most cases showed epilepsy first before the age of three years and that it continued in the majority. All the children in whom epileptic attacks ceased to occur had fits more frequently than once a month during the time it had been evident. None of them showed petit mal.

The severity of grand mal attacks did not seem to have any relationship to the frequency of their occurrence in the small number of cases in the series.

Jacksonian attacks were shown by two cases, one triplegic and one tetraplegic. In each case they began in the upper limb /

TABLE 129

Strabismus in 79 diplegic patients

	No. of cases	Oculomotor weakness		Abducent weakness		Total number of patients
		Unilateral	Bilateral	Unilateral	Bilateral	
Paraplegia	79	1	0	4	4	9
Triplegia	23	0	1	4	6	11
Tetraplegia	27	3 *	3	3 *	6	14
Diplegia	79	4 *	4	11 *	16	34

* one case showed associated unilateral oculomotor and bilateral abducent paresis.

TABLE 130

Epilepsy in 79 diplegic patients

	No. of cases	Type of attack			Myoclonic attacks	No. of patients
		Grand mal	Jacksonian	Petit mal		
Paraplegia	29	6	0	1	0	6
Triplegia	23	8	1	3	0	8
Tetraplegia	27	6	1	2	3	7
Diplegia	79	20	2	6	3	21 *

* 8 had attacks of more than one type

TABLE 131

The age at the time of the first fit and the last fit in grand mal and petit mal occurring in 79 diplegic patients.

		Under 1 year	Over 1 year and under 3	Over 3 years and under 6	Over 6	Present at time of examination
Grand mal	First fit	7	5	6	2	-
	Last fit	0	1	3	0	16
Petit mal	First fit	2	0	2	2	-
	Last fit	0	0	1	0	5

TABLE 132

The frequency of attacks of grand mal in 79 cases of diplegia.

More than one attack in a week	2
Less than one attack in a week More than one attack in a month	10
Less than one attack in a month More than one attack in a year	6
Less than one attack in a year	2
No. of patients with grand mal	20

limb of the more affected side. These patients were extremely interesting in that they tended to show more marked facial paresis on the affected side, and the limbs on that side were less useful following attacks. Immediately after attacks the clinical picture was very similar to that of hemiplegic paresis, with neglect of the affected limbs and an accentuation of the spastic signs on the affected side, except that the contralateral lower limbs were also involved more or less severely.

Petit mal occurred in six patients, all of whom showed grand mal in addition. The attacks were present at least once daily in all these cases, and in two occurred more frequently than six times a day.

The cases classified as having myoclonic jerks were all cases of moderately severe or severe tetraplegia. In all the attacks were initially associated with dystonic attacks. In one the attacks persisted after dystonic attacks had ceased to occur, in the other two the attacks ceased with the attacks of dystonia. In the latter two cases the attacks began with dystonic movements, usually on handling, similar to those which have been described earlier. The child became stiff, the back, head and limbs extended; but not only was consciousness impaired slightly, but it appeared to be very severely impaired and the child immediately became limp and remained so for a few seconds before returning to normal. In the third case the attacks began similarly, but the child seemed actually to lose consciousness at the time of the movement, and the attack /

attack was described as being "head nodding" in type.

It is interesting to note that the three children showing myoclonic jerks all showed other epileptic manifestations, grand mal and petit mal in one, grand mal alone in one, and petit mal alone in one.

The success of therapy in epilepsy associated with diplegia appears to be rather poor. Though the frequency of attacks, whether grand mal or petit mal, appears to be reduced, only four cases ceased to have attacks while on therapy, and there is nothing to indicate that this was a direct result of therapy.

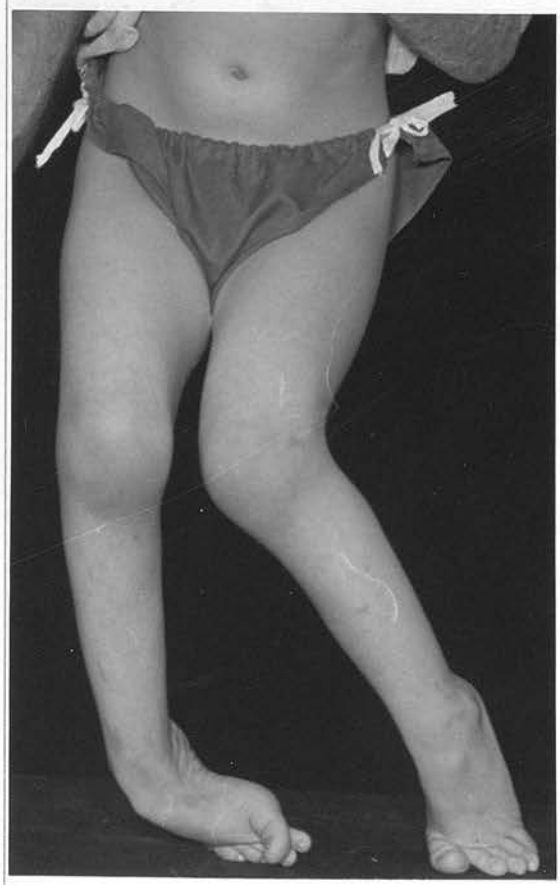
Other Skeletal Abnormalities in Diplegia.

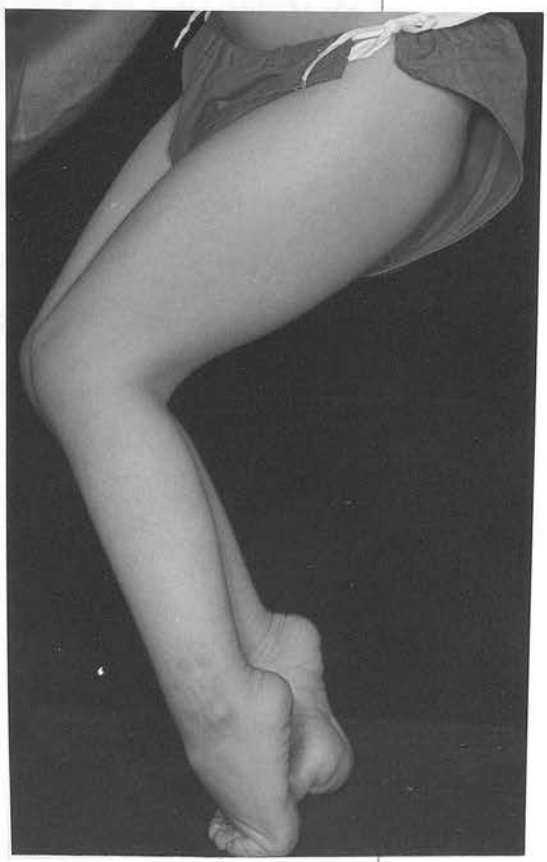
Dislocation of the hip was encountered in four patients with severe paresis. Three of these were tetraplegic and one triplegic. In one case of tetraplegia the dislocation was bilateral. The dislocation appeared in one between two examinations, between which the degree of rigidity of the lower limbs had very markedly increased. The acetabulum was noted to be poorly formed in two cases, but it seems quite clear that the dislocation is secondary to the extremely powerful pull of the hamstrings in diplegia. Since the survey was completed it has become apparent that secondary dislocation of the hip is much commoner amongst diplegic patients than had been suspected.

Two cases of diplegia were encountered in which arthrogryposis was present, moderately severely in one and slightly in another. It is interesting to correlate the common positions /

Figure 14.

Illustrating the marked deformities of the lower limbs which may occur in severe diplegia in the stage of flexion contracture.(case 59).





positions assumed by the limbs in arthrogryposis to those in severe tetraplegic paresis.

A degree of talipes equino varus, evident at birth and not associated with arthrogryposis, was noted in three cases of diplegia. One case was paraplegic and two were triplegic. In all the cases the talipes was bilateral, though asymmetrical in severity.

As in hemiplegia, it seems probable that there is some aetiological connection between congenital talipes equino varus deformity and diplegia (Freud, 1897).

Other slight abnormalities of the limbs were found. Syndactyly of the second and third toes occurred in a number of diplegic patients. The hands and feet appeared to be abnormally small in a high proportion, the fingers tending to be abnormally equal in length, soft, hyperextensible, and poorly formed.

Other Abnormalities.

Congenital heart lesions were encountered in two patients. One was though to have a patent intra-auricular septum, and one a patent intra-ventricular septum.

Naevi were encountered in nine patients, a finding of interest in view of the recent work on the relationship between haemangiomata, congenital eye defects, and cerebral abnormalities (Ingalls, 1943).

One patient with familial history of neurofibromatosis showed a rather muddy yellow skin with patches of brown pigmentation.

Congenital /

Congenital Anomalies.

As has been described, a number of patients had congenital anomalies. Some of these were of minor importance. Twelve, for example, had hyperextensibility of the fingers, associated with high inter-digital webs, digits of rather equal length and small size. In 6 of these there was partial syndactyly of two or more digits, most commonly the second and third, in feet and hands (Table 133).

In a further 13 cases similar anomalies of the digits were accompanied by other congenital abnormalities, and in 5 other abnormalities occurred alone. As will be seen from the Table, talipes equino varus was the commonest of these, occurring in a total of 6 cases, while congenital heart disease occurred in three, multiple naevi in three, arthrogryposis in two and hyperteleorism in one.

Since there may legitimately be some dubiety about the significance of hyperextensibility of the digits and slight syndactyly of the digits in a bilateral neurological disorder, the percentages of cases with other anomalies have been estimated as well as the total with hyperextensibility and syndactyly included.

The proportion of cases with congenital anomalies who had other abnormal siblings, abnormal parents or abnormal relatives was not significantly different from those cases of diplegia without anomalies.

TABLE 133

Other congenital anomalies in patients with congenital diplegia.

Description of anomaly.		No. of patients	% of patients
Hyperextensibility of the digits only.		6	8
Hyperextensibility of digits and partial syndactyly.		6	8
Hyperextensibility of the digits, partial syndactyly and other anomalies		13	17
Nature of these other anomalies			
Hypomandibulosis and high arched palate.	3		
Talipes equino varus.	5		
Arthrogryposis.	1		
Congenital heart disease.	2		
Multiple naevi.	1		
Hyperteleorism.	1		
Multiple naevi only.		2	
Congenital heart disease only.		1	
Talipes equino varus deformities only.		1	
Arthrogryposis only.		1	
Total		30	39
Total excluding those with hyperextensibility of digits and syndactyly only.		18	23

Review of the literature on ataxia and ataxic diplegia.

Interest in children suffering from ataxia was stimulated by the description of cases of progressive ataxia by Friedreich, (1861, 1863, 1876). Though there was a good deal of scepticism, especially amongst French authors, about this being a clinical syndrome distinct from disseminated sclerosis, the description of further cases and the acceptance of Friedreich's cases by Gowers, stimulated further research, (Charcot, 1887; Gowers,1880). Freud described two patients suffering from "diplegia" in whom there was intention tremor, mystagmus and ataxia but he did not consider these signs sufficiently important to make a special category of cerebral palsy for them. He was more concerned with distinguishing them clinidally from cases of progressive hereditary ataxia, (Freud, 1893). Later he discussed the possibility of finding sufficient cases of non-progressive ataxia in childhood to consider them a clinical entity within the category of "cerebral palsy", (Freud, 1897).

Congenital ataxia. Eight cases of congenital ataxia ("cerebellar diplegia") were described by Batten only six years after Freud's suggestion. In at least three of them there was a history of abnormal labour or delivery. Retarded motor milestones, defective speech and gross unsteadiness and cluminess were the presenting symptoms. On examination there was marked unsteadiness, incoordination of the limbs, intention tremor and sometimes nystagmus. All but one of the eight cases were hypotonic and in some the lack of

muscular /

muscular tone was striking though the tendon jerks were usually brisk. In the exception, a girl first seen at the age of seventeen months there was well marked rigidity of the lower limbs, and ankle clonus with extensor plantar responses were found when she was re-examined at the age of eight years. She remained unsteady. Batten stressed that there was a tendency for patients suffering from congenital ataxia to improve as they matured and noted that independent walking was almost always accomplished. He thought the condition could be caused by damage to the cerebellum during the birth process, or by cerebellar hypoplasia, some of his analogous cases being kittens with cerebellar hypoplasia delivered after normal labour" to a half-bred Persian belonging to "the Sister of Hope Ward in St. Bartholomew's Hospital". In passing, he mentioned that "diphtheretic paralysis, acute vascular lesions of the cerebellum, cerebellar tumours and abscess may all give rise to ataxia of the cerebellar type". He distinguished these conditions and the progressive hereditary ataxias largely by the different histories and clinical courses of the patients. Two years later he described two cases of congenital ataxia in more detail and distinguished them from "Cases in which ataxia has suddenly developed after some acute illness in a child who formerly has been quite healthy, (acute ataxia, encephalitis cerebelli)" and cases of hereditary ataxia. He described cases of acute ataxia following /

following whooping cough, measles and scarlet fever, (Batten, 1903, 1905).

In these two papers, Batten succeeded in showing that clinical syndromes of congenital ataxia, congenital ataxia diplegia, acquired ataxia and acquired ataxia with spasticity in the limbs, could be distinguished.

Descriptions of four more cases of cerebellar ataxia were given by Förster, (1909). These cases differed somewhat in their clinical findings from those previously presented. The main characteristic was an extensive hypotonia with undue passive mobility of the joints. All four were mentally deficient and two were completely mute at the age of three, whilst the other two who were older had defective articulation. They were quite unable to maintain posture, but could move with normal power, though rather jerkily when lying down. There was incoordination of agonists and antagonists in movement. Tremor was present. Jerks were sluggish and plantar responses flexor. In two cases which came to autopsy sclerosis of the frontal lobes was the only striking pathological abnormality but it is noteworth that in neither case the cerebellum was examined microscopically. Förster was at some pains to distinguish the findings in these patients from those of cerebral diplegia in which increase of tone was present in the limbs. It is of great interest, however, to note that one of his younger patients whom he considered to show an atypical example of his syndrome, was liable /

liable to assume positions of opisthotonos when handled, (and sounds suspiciously like a case of diplegia in the dystonic stage). Another of his cases in which there was spasticity in the lower limbs and extensor plantar responses whom he thought showed a transitional form between congenital ataxia and congenital cerebral diplegia would undoubtedly be classified as suffering from ataxic diplegia in the present classification.

Further series of cases similar to these described by Forster were soon forthcoming. "Infantile cerebrocerebellar diplegia of flaccid ataxic astasic type" was stated to be fairly common by Clark, (1918). He added little new to the clinical picture which had previously been presented. Four patients with many of the clinical features described by Forster, but in whom hypotonia was less extreme were presented by Batten and von Wysse, (1915). These authors noted that hypertonus was actually present in many cases when the patients were placed upright. A patient who showed similar features on clinical examination to those described by Batten and von Wysse was demonstrated to the Royal Society for Medicine in 1916 by Parkes Weber, (1917).

Four patients with the "ataxic astasic" type of diplegia were described by Hunt, (1918). All four had a history suggestive of birth injury which Clarke had stated to be unusual. All had retarded motor and speech milestones and though there was no paralysis or spasticity, /

spasticity, coordination of the limbs was much impaired. Marked hypotonia was present and all showed intention tremor whenever they started to move. None was mentally defective, though speech was impaired in all. He felt the prognosis was much better than in "Ataxic Diplegia" in which there existed "a very grave form of birth palsy". In fact, his cases appear to be more like those of Batten and von Wysse than those of Forster.

The descriptions of later authors have contributed rather little to the pictures of congenital ataxia which had been presented earlier. Cases similar to those of Batt en have been described briefly by Phelps, (1940) and Perlstein, (1952). Cases similar to those of Forster have been described and discussed by Ford,(1952). The differential diagnosis of "Atonic Cerebral Palsy" has been discussed by Horton and Yannet, (1952). These workers present cases whose clinical pictures vary from those similar to that defined by Forster, to that described by Batten. The tendency for their to be intertypes between these pictures has also been noted by Kinnier Wilson, (1954).

Unfortunately relatively little pathological corroboration of the suggestions made as to what may be the underlying cerebellar pathology has been forthcoming. There seems little doubt that a proportion of cases is due to birth injury and a proportion to developmental abnormalities of the cerebellar system complex. A striking example of cerebellar hypoplasia resulting in congenital /

congenital ataxia has been presented by Baker and Granit, (1931). But it is indicative of the poor correlation of clinical and the pathological observations so far achieved that no clinical histories of patients with cerebellar agenesis or hypoplasia are presented in the monograph of Brouer and Biemond, (1938). Perhaps the more frequent diagnosis of congenital cerebellar ataxia may result eventually in an improvement in this state of affairs.

The extent to which ataxic diplegia has been ignored by modern authors is striking, for the existence of cases showing marked spasticity in the lower limbs in association with cerebellar ataxia has been recognised from the time of Batten, (1903). Brief reference to cases coming into this category has been made by Stewart,(1942) Penrose,(1938; and Yannet,(1949). But detailed clinical descriptions are not presented and the cases are grouped as "Diplegic patients with ataxic signs". More importance is given to the category by Kinner Wilson, (1954). He calls it "Cerebro cerebellar diplegia". There is great need for a large series of fully described cases in order that the clinical picture can be further defined and the importance of heredity in the condition established.

Hydrocephalus. Though hydrocephalus has always attracted curiosity as a phenomenon of nature, only when the /

the possibilities of surgical attack began to be explored was an era of scientific investigation initiated. Following the pioneer work of Dandy and Weed in the United States, and Dott in this country, the different clinical and pathological forms of hydrocephalus began to be distinguished. For example, a classification based on probable aetiology and site of obstruction was presented by Fraser and Dott, and operations described for dealing with the commoner types in infants who were not already too severely damaged, (Dandy and Blackfan, 1913,1914; Dandy, 1917, 1919; Fraser and Dott, 1922; Kausch, 1908).

Work since this time had led to considerably fuller underst anding of the pathology of hydrocephalus and the recognition that it may be the result of a very wide variety of causes. Russell lists maldevelopments, including deformities of the aqueduct, spinal bifida and deformities of the base of the skull, gliosis of the aqueduct of uncertain origin, inflammations, especially meningitis, dural sinus thrombosis and thrombophlebitis and neoplasms, (Russell, 1949).

Of particular importance from the point of view of cerebral palsy in childhood are the forms of hydrocephalus which become arrested spontaneously and those which are now susceptible to forms of neurosurgical treatment. In the latter category are an increasing number of children with aqueduct stenosis or atresia and those with meningeal adhesions in the posterior fossa due to fibrosis /

fibrosis following haemorrhage at the time of birth or the result of meningitis. In spite of surgical alleviation of their hydrocephalus or its spontaneous arrest, a proportion of such patients show permanent neurological defects. According to Ford, patients with meningeal adhesions in the posterior fossa or involving the roof of the fourth ventricle are particularly liable to show ataxia, (Ford 1952). Unfortunately insufficient numbers of patients have so far been reported to confirm this impression from other sources. It is already apparent, however, that the common permanent neurological defects encountered are ataxia and spastic paresis, usually more severe in the lower limbs than the upper. Thus, a clinical picture of ataxia or ataxic diplegia may present, occasionally in association with optic atrophy, hemiplegia and, especially after meningitis, deafness and vestibular derangements. The degree of mental impairment appears to be very variable, but difficulties in the organisations of spatial relationships are common and frequently lead to very great difficulty in learning to read and write, (Dunsdon, 1953).

Acquired Ataxia. The occurrence of ataxia complicating infectious diseases was well known in the second half of the last century. The majority of reported cases recovered completely within a few weeks or months. But children who showed persistent ataxia with or without associated /

associated spasticity following infections were also described after scarlet fever by Shepherd, (1868), Voelcker, (1905), after measles by Batten, (1905), Clapton, (1871), Guthrie, (1905), after whooping cough by Hammarburg, (1890), Gordon, (1918), after dysentery and typhoid by Feith, (1873), Lenhartz, (1 883), Marie and Joltrain, (1910). Ataxia complicating diphtheria appears to have been due to sensory neuropathy rather than cerebellar damage in most instances, but occasional cases of cerebellar ataxia were noted, (Wilson, 1919; Dolgopol and Katz, 1950). Similar cases of acute ataxia which persisted complicating upper respiratory infections, influenza and other ill-defined illnesses were also reported, (Batten, 1907; Bastian, 1878, Taylor, 1904). In other cases the ataxia came on without any preceding illness being apparent, (Seham, 1919). The cases of persistent ataxia which these authors described were greatly outnumbered, however, by those in which complete recovery ensued, (Griffith, 1916, 1921).

Other less frequently discussed causes of persistent ataxia were those in which cerebellar damage was due to trauma and those in which septic intracranial conditions were responsible. Thus children who were made permanently ataxic as a result of intracranial abscess, or lateral sinus thrombosis, most often complicating middle ear disease were reported by Gowers, (1899), Sachs, (1926), ~~Ballome, (1908)~~. /

~~Ballome, (1908)~~. A full review of these conditions has recently been presented by Brunner, (1946).

Unfortunately very few systematic reviews of the long term effects of acute septic meningitis on the neurological state of those children who survived it are to be found in the literature. Occasional case reports of children who showed persistent ataxia following meningitis are available, and it is apparent that even in the pre-chemotherapeutic and pre-antibiotic era, ataxia was one of the residual effects which might occasionally be expected, (Wilson, 1939). Systematic reviews of the neurological defects encountered are few. The rarity of reports of patients who showed persistent ataxia may be the result of ataxia being the most difficult form of cerebral palsy to diagnose in childhood. But it is nevertheless disappointing to discover so few cases recorded, (Perlstein, 1952; Ingram, 1955). In this connection it is interesting that the author has seen at least four cases in the past four years who showed moderately severe cerebella/ataxia who were classified as being completely cured of tuberculous meningitis in a published review of the effects of streptomycin and other ancillary treatment. In the experience of many clinicians who see large numbers of patients with cerebral palsied children, ataxia following septic or tuberculous meningitis is not too uncommon. Yet a review of the recent literature presenting surveys of patients having recovered from this condition shows a surprising lack of awareness that this complication exists /

exists. For example, hydrocephalus and ataxia are mentioned, but no exact figures of the frequency of these complications are given in his follow-up of 409 cases of septic meningitis by Smith, (1945), or in 169 cases by Maltke and Roaschau and Nicolson, (1954). It is impossible to assess how frequently ataxia occurred as a result of tuberculous meningitis and its treatment in the cases reported by Daelen, (1953), Riser, Geraud and Ribaut, (1954), Oldham et al, (1954), and Robinson and Ro (1953). Yet cerebellar or vestibular ataxia with or without associated spastic paresis in the limbs is often found in patients who have been treated for tuberculous meningitis, and who come for treatment to clinics for handicapped children. That they should appear so frequently is something of a corrective to placing too much trust in published results in which follow-up of patients is often inadequate.

It is convenient briefly to review the more recent literature describing cases of persistent ataxia in childhood at this point. In order not to complicate description unnecessarily, no distinction is made between cases of persistent ataxia in which there was associated spastic paresis of the limbs and those in which this was absent. Some of the recorded cases would be classified as suffering from ataxia and some from acquired ataxic diplegia. /

diplegia.

Acute disseminated encephalomyelitis. (Acute perivascular demyelinisation). A large number of reports of acute ataxia complicating measles, varicella, rubella and ill-defined infections are available in the literature. Most children who suffered from ataxia as a result of acute disseminated encephalomyelitis recovered completely within a few weeks, (Miller and Stanton, 1956). But cases of persistent ataxia following measles are reported by Marsh, (1910), Guthrie, (1905), Skoog, (1920), Ford, (1928) and Griffith, (1952). Miller and Stanton state that three of 224 cases of measles encephalomyelitis which they found in the literature appeared to show persistent ataxia. Cases following chickenpox were reported by Grenet and Georges, (1938), Dagnelie and Dubois, (1932), Applebaum and Rachelson, (1953). Miller and Stanton note that acute ataxia complicating varicella occurred in about 16% of reported cases with neurological complications and persistent ataxia appears to be uncommon. This point is also stressed in his comprehensive review of the neurological complications of ~~vacidla~~ varicella by Vydevard (1935). A case of persistent ataxia complicating rubella was reported by Margolis et al, (1943). Miller and Stanton also refer to a single case following mumps encephalomyelitis. It has proved impossible to find a well authenticated case with adequate /

adequate follow-up of persistent ataxia following vaccinial encephalomyelitis who showed persistent ataxia though there seems no good reason for assuming that chronic ataxia should not result from the condition, (Turnbull and Mackintosh, 1926). Persistent ataxia following acute disseminated encephalomyelitis without a well defined pause was reported by McAlpine, (1929.) Case 7 of Redlich (1927) also seems to fall into this category.

Toxic encephalitis. Persistent ataxia resulting from toxic encephalitis complicating other infections have been noted. Diphtheria was observed to be a cause in patients reported by "Serog", (1916); Bruckner, (1909); Dolgopol and Katz, (1950). In his discussion of Bruckner's paper, Rostowski notes, "Weist daran, hin, dass man schwere Ataxie nach Diphtherie öfters finde ....." which suggests that it occurred not infrequently !!!

Persistent ataxia following scarlet fever was noted by Raymond and Rae (1906) and Lassen and Bang, (1940). Miller and Stanton note that ataxia was a feature of 36.3% of cases of scarlet fever with intracerebral complications reported in the literature. The ataxia was rather seldom persistent. Similarly though there are a number of reports of acute transient ataxia occurring during the course of pertussis, usually after the second week, the only recent cases of persistent ataxia noted are /

are those recorded by Kokken, (1940), Litvak et al, 1 948), and Griffith, (1952).

The literature describing persistent ataxia in childhood resulting from cranial trauma appears to be scanty, though there can be no doubt from clinical practice that such cases occur. Similarly persistent ataxia may follow successful exploration of the posterior cranial fossa for the removal of tumours or treatment of inflammatory lesions.

Recent Surveys.

Recent surveys of large series of patients suffering from cerebral palsy have not added a great deal to what was previously known about the clinical findings in ataxia and ataxic diplegia. This is partly because p atients suffering from ataxic diplegia have been grouped together either with those with ataxia or those suffering from symmetrical spastic disorders, (Asher and Schonell, 1950; Woods, 1957).

The proportion of patients considered to be ataxic varies greatly in different series. One per cent. of patients suffering from cerebral palsy were classified as suffering from Asher and Schonell, (1950), and Floyer, (1954). Phelps and Perlstein suggest that a figure of about 5% represents the proportion of ataxic patients, (Perlstein, 1955). But higher figures have been reported in /

in other surveys. Andersen and Woods quote 10%, (Andersen, 1954; Woods, 1957). In the latter series, seven of the 49 cases classified as ataxia appear to have been suffering from progressive hereditary ataxia and would not be considered to suffer from cerebral palsy by most authors. The corrected proportion of ataxic patients is about 7%. This figure is similar to that given by Skatvedt, (1958).

The intelligence of patients with ataxia has been studied in a number of series, and estimates differ considerably. Thus Woods, (who included her progressive cases) found that 72% were of "normal" or superior intelligence, and that only 28% had intelligence quotients below 70. On the other hand, 62% were found to be less intelligent than this by Asher and Schonell, (1950). Skatvedt found that 10 of 18 patients were of average intelligence, 6 had intelligence quotients between 50 and 85, and two quotients of less than 50, (Skatvedt, 1958). The great difficulties encountered by some ataxic patients in direction finding and in educational work involving spatial concepts have been well described by Dunsdon, (1952).

Epilepsy is less commonly encountered amongst ataxic than amongst diplegic patients according to most authors. 27% of her mixed group of patients were noted to be epileptic by Woods, (1957), and 17% by Skatvedt, (1958). This /

This author noted grand mal convulsions and myoclonic seizures in her patients. No focal seizures were observed. The electroencephalogram showed diffuse dysrrhythmia more frequently than focal features, a finding confirmed by Anderson, (1954).

Few systematic studies of hearing or speech defects have been made in ataxic patients. Two of eighteen patients had impaired hearing when tested by audiometry, and defects of vision, hearing and speech were noted by Skatvedt. Three of her eighteen patients suffered from strabismus, two from refractive errors, and two from impaired hearing. Nine of her patients were dysarthric, and in others there was retardation of speech development. Speech defects were noted to be commoner amongst ataxic patients than amongst those suffering from bilateral spastic paralysis, (Skatvedt, 1958).

CHAPTER 8b

Clinical Study of patients suffering from Ataxic Diplegia.

Family history of patients with ataxic diplegia.

In Table 134 are shown the details known about the neurological, mental and psychiatric disorders occurring in the families of patients with ataxic diplegia. Both parents were apparently free of disorders of this type in eight cases. In two cases only, one parent was abnormal. (The father in Case 27 suffered from anxiety neurosis and a duodenal ulcer) which had resulted in admission to mental hospitals on a number of occasions. In Case 101. the mother was feeble minded, had obvious healed rickets

TABLE 134

Family history of patients suffering from ataxic diplegia

Case No.	Social Class	Health of father	Health of mother	Health of siblings	Health of other relatives	Other miscarr. & stillbirths	Consang-uinity.
8	III	Good.	Good.	1 Health.	Health.	2 neonatal deaths erythroblastosis.	No
14	III	Bronchitis.	Good.	One ½ sister had breath-holding conv. the other ½ sister has hemi-plegia.	Brother of father backward.	None.	No
16	II	Anxiety neurosis.	Pes cavus.	3 healthy.	Healthy.	None.	No
27	III	Neurasthenia. Duodenal ulcer.	Healthy.	1 healthy.	Healthy.	None.	No
57	V	Alcoholic and ment. unstable.	Mild ataxic diplegia. F. minded.	1 died Tub.Meng.	Mat.uncle defect. Mat.G.mother sev-ere depression.	One stillbirth.	?
87	III	Healthy.	Healthy.	-	Healthy.	None.	No
91	III	Healthy.	Healthy.	-	Healthy.	One abortion.	No.
101	V	Died of cancer.	Feeble minded. Healed rickets. Spastic in legs.	1 healthy.	Healthy.	None.	No.
135	IV	Healthy.	Healthy.	2 healthy.	Healthy.	None.	No.
150	IV	Healthy.	Healthy.	7 healthy.	Healthy.	None.	No.
156	V	Healed tub.	Healthy.	2 healthy.	Mat.uncle hydro-cephalus & epil. 2 of father's sisters epil.	None.	No.
182	III	Healthy.	Healthy.	1 healthy.	Healthy.	Stillborn twins.	No.

rickets and was unsteady on her legs and walked on her toes. Her tendon jerks were exaggerated and her plantar responses were extensor. She stated she had been slow in walking and talking and had gone to a special school. She had always fallen frequently. It was considered that the probable diagnosis was ataxic diplegia in her case.

In 2 cases, both parents were abnormal. In Case 16, the father suffered from anxiety neurosis. The mother had marked pes cavus, similar to that observed in her daughter who had become ataxic between the ages of 5 and 7. The mother showed no evidence of other neurological disease, however.

In Case 57 the mentally unstable and alcoholic father had left home. The mother was a feeble minded prostitute who had always been unsteady according to her mother and still wore out the toes of her shoes. Like the younger sister of the patient, it was felt likely that the mother suffered from mild ataxic diplegia.

Siblings. There were 22 siblings of whom details were known. Not all of them could be examined fully, (most regrettably the ~~younger~~ older sister in Case 57 who is not, therefore, included with the patients diagnosed as ataxic diplegia in this series). One child died from tuberculous meningitis, another younger sister of Case 57.

In /

In Case 14, one half-sister (the mother's children by a previous marriage) had convulsions associated with breath-holding attacks until the age of four, but was otherwise well. Another had a left hemiplegia apparently of congenital origin. She was over 14 at the time of the survey.

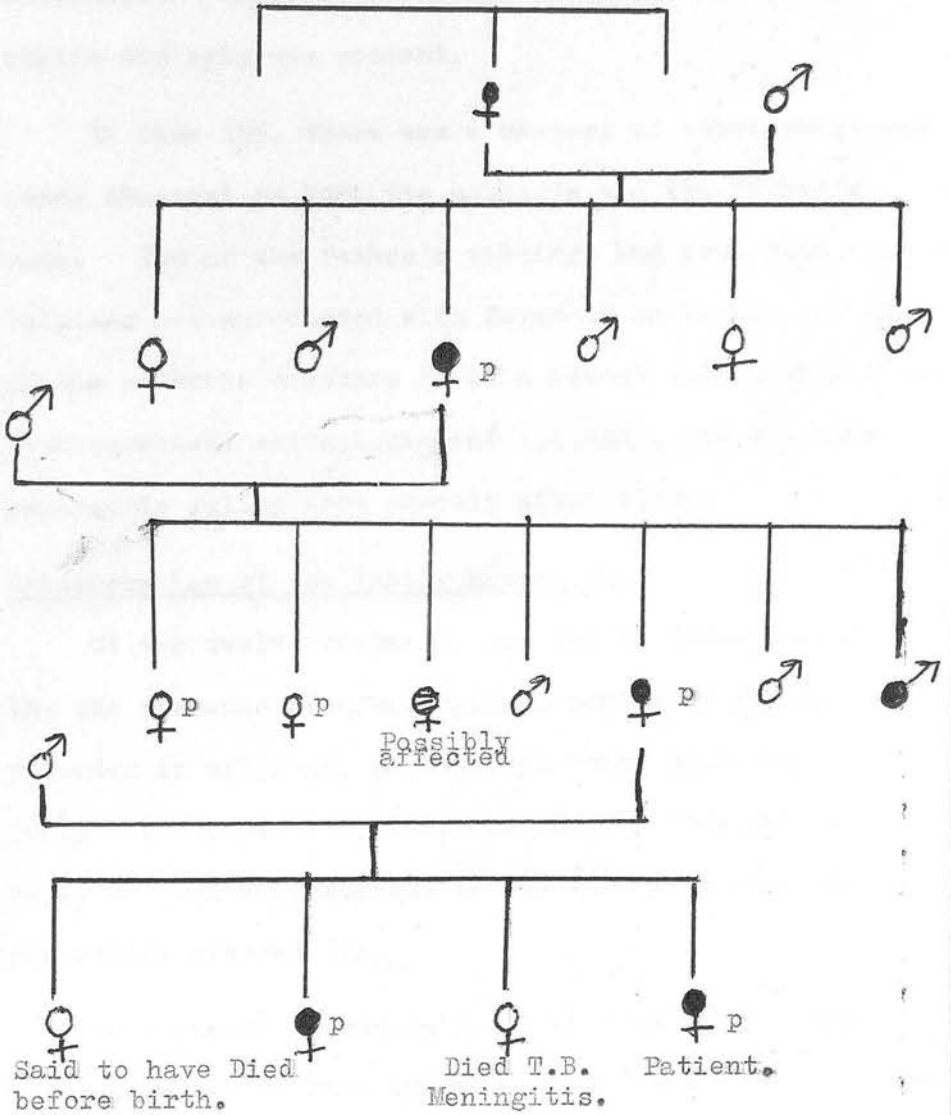
The remaining 17 siblings were healthy ..... approximately 77% of twenty-two.

Other relatives. No other relatives were known to be abnormal in 9 cases. In Case 14, one brother of the father was mentally retarded, said his first words at the age of over four, and in spite of going to a special school, had made little progress in learning to read or write.

In Case 57, the maternal grandmother stated that she herself, her mother and three of her six children, including the mother of the patient had always walked on their toes and been unsteady. The most severely affected was the mother's brother who was mentally defective and was traced to an institution for mental defectives where he was diagnosed as "spastic paraplegia with some hand involvement". Permission to examine him w as refused. Unfortunately the mother's other affected sibling, a sister, could not be traced. There seems little reason to doubt that she did suffer from ataxic diplegia, however.

The /

The Family History in Case 57.



Known to be affected.

Possibly affected.

Personally examined. p

The grandmother and the mother were examined as far as was compatible with their refusal to undress. They were both unsteady on their feet, walked on broad bases, were clumsy when handling objects and had increased tendon jerks in the lower limbs and extensor plantar responses. There seems little doubt that familial ataxic diplegia was present.

In Case 156, there was a history of other relatives being abnormal on both the mother's and the father's side. Two of the father's siblings had recurrent convulsions not associated with fever as children, and one of the mother's brothers is in a mental institution with hydrocephalus, tetraplegia and epilepsy, the symptoms apparently dating from shortly after birth.

Consideration of the family histories.

Of the twelve patients, six had no history suggesting the presence of neurological, mental or psychiatric disorder in siblings, parents, or other near relatives. (Cases 8, 87, 91, 135, 150, and 182). In a further case, 27, anxiety neurosis in the father was the only presenting abnormality.

In 5 cases, disorders to which more significance might be attached were present. In 3 cases, the mother showed important traits, ataxic diplegia in Cases 101 and 57, and pes cavus in Case 16. In these cases it seems probable that a dominant inheritance of the ataxic diplegia /

diplegia was occurring.

In 2 other cases, 14 and 156, it is more difficult to assess the significance of family history, but epilepsy was apparently present on both sides of the family in Case 156, though it was probably idiopathic on the father's side and "organic" in the case of the mother's brother.

History of the other pregnancies of the mothers of patients with ataxic diplegia.

Including the pregnancies resulting in the birth of the patients there were 41 conceptions to the mothers of the twelve patients with ataxic diplegia. This gives a fertility of 3.4 pregnancies per mother, compared to 2.8 for the mothers of diplegic patients, and 3.5 for the mothers of all hemiplegic patients (2.8 for mothers of patients with congenital hemiplegia, and 3.8 for mothers of children with acquired hemiplegia).

Of the 29 other pregnancies to these twelve mothers, five ended in abortion, one in Cases 27, 87, 91 and 2 in Case 156. Two ended in stillbirth, the cause being unknown in Case 57, and attributed to preeclampsia in Case 182, in whom the pregnancy was twin and both twins were dead on delivery.

In Case 8, in which the patient was the first-born, the second child was midly affected by rhesus incompatibility, and the two succeeding children died shortly after delivery from severe erythroblastosis, the last born being hydropic. In Case 57, there was one infant death /

death from tuberculous meningitis at the age of 4 months. Thus, only 19 children were alive at the end of the year as a result of the 29 other pregnancies, and as has been noted, three of these survivors were abnormal. Only 54% of the other pregnancies resulted in the delivery of healthy children who survived the first year. (Table 134)

As indicated in the table, the common actual abnormalities of parturition in these pregnancies were abortion, preeclampsia and rhesus incompatability. Abnormal delivery occurred three times, one being precipitate spontaneous, one being an assisted breech, and one forceps. It is quite probable that other labours were also terminated by forceps, for when these occurred at home the mothers frequently did not know whether instruments had been used or not. Including abortions, but otherwise ignoring outcome, eleven of the twenty-nine other pregnancies, 41% were abnormal in some respect.

Spacing of pregnancies. Though the number of cases is too small for any formal statistical evaluation, it will be seen from Table 135 that the spacing of pregnancies shows the same trend that was noted in both congenital hemiplegia and in diplegia. The lapse of time between the last pregnancy prior to the birth of the patient and the birth of the patient and the succeeding pregnancy is greater than the lapse of time between subsequent or preceding siblings.

Place in the family and number of pregnancy. /

Place in the family and number of pregnancy. As shown in Table 135 the observed distribution of patients with ataxic diplegia by place in family and number of pregnancy does not differ significantly from the expected distribution.

TABLE 135

Maternal age.

Maternal age	Number of patients	Congenital	Acquired & hereditary
Under 20	0	0	0
20-25	3	1	2
25-30	4	3	1
30-35	2	2	0
35-40	3	1	2
40 or more	0	0	0

The distribution of cases by the maternal age at the time of birth of the patients is shown in the above table. The small number of patients makes it impossible to draw any conclusion about the significance of maternal age in ataxic diplegia, but it does suggest that the distribution is not startlingly different from the average for the general population.

Maternal health.

Apart from the conditions which have been referred to under the heading of family history, the mothers of patients with ataxic diplegia, on contrast to those with diplegia /

diplegia appear to be remarkably healthy. In particular, there is no reference to gynaecological disorders and (by a rather odd chance) full details of the menstrual histories in eight of the women were available. All were normal.

On the other hand, it must be remembered that at least two of the mothers were very unintelligent, and living in very poor social conditions (Cases 57 and 101). Their accounts of their health records may not be very reliable.

The birth histories of patients with ataxic diplegia.

In Table 136 are summarised the clinical details of the birth histories of the twelve patients with ataxic diplegia. In three cases there were no apparent abnormalities in pregnancy, labour, or delivery, and the infants seemed well immediately after birth, (Cases 27, 182 and 57. In Case 16 the mother had varicose ulceration of the legs during pregnancy as the only abnormality and since pregnancy and delivery were otherwise uncomplicated this case will be considered in the "normal" group.

In two cases, 8 and 150, pregnancy only was abnormal. In the former there was a small vaginal haemorrhage 12 days before the onset of labour which was at an estimated 36 weeks. Since the mother had a funnel shaped pelvis this premature onset of labour may have been quite convenient for the child was delivered spontaneously by the vertex in good condition after a labour of 16½ hours. Her birth weight was 5 lbs. 1 oz. In the second case, labour was induced by quinine by mouth and pituitrine injections at an estimated 41 weeks gestation because the mother was becoming so depressed. Though the five-hour labour seemed uncomplicated the child was born intensely cyanosed, apnoeic and was very slow to respond to resuscitation. Generalised convulsions occurred on the third day of life.

In only Case 135 was labour or delivery abnormal.

In /

TABLE 136 A

Birth histories of patients with acquired ataxic diplegia.

Case No.	Social Class	Mat. Age	Number of pregnancy	Maternal health	Health in pregnancy	Duration of labour	Mode of delivery	Birth weight	Condition of baby	Placenta
87	III	23	1st. and only.	Good.	Mild pre-eclampsia	14 hrs.	Mid cavity forceps.	8/3¼	Good.	Normal.
182	III	28	2nd. of 3	Healthy.	Healthy.	2 hrs.	Spont.vertex deliv.	10/-	Good.	No note.

TABLE 136 B

Birth histories of patients with probable hereditary ataxic diplegia

Case No.	Social Class	Mat. Age	Number of pregnancy	Maternal health	Health in pregnancy	Duration of labour	Mode of delivery	Birth Weight	Condition of baby	Placenta
16	II	39	4th. of 4	Pes cavus. Otherwise well.	Varicose ulcers of legs.	9 hrs.	Spont. vertex.	7/-	Seemed normal.	No information.
57	V	23	4th. of 4	Ataxic diplegia.	Healthy.	5 hrs.	Spont. vertex.	6/3	Normal.	Unknown.
101	V	38	2nd. of 2	Ataxic diplegia.	Good but premature onset of labour.	1 hr.	Precipitate spont. deliv. by vertex.	3/8	Fair. Premature.	Unknown.

TABLE 136 C

Bith histories of patients whose ataxic diplegia was thought to be congenital but not hereditary

Case No.	Social Class	Mat. Age	Number of pregnancy	Maternal health	Health in pregnancy	Duration of labour	Mode of delivery	Birth weight	Condition of baby	Placenta
8	III	23	1st. of 4	Good. Funnel shaped pelvis.	Small APH 12 days before labour.	16¾ hrs.	Spont. vertex.	5/1	Normal. Would not fix to breast initially.	1 lb.
14	III	38	3rd. of 3	Well.	Haemorrhage at 11 weeks. ? abortion.	4 hrs. sev. pain.	Difficult mid cavity forceps.	6/12	White asphysia. Coramine. Artif. respiration.	No note.
27	III	31	2nd. of 2	Good.	Healthy.	8/25	Spont.vertex with episiotomy.	9/2	Good.	Healthy. Post partum haemorrhage.
91	III	26	2nd. of 2	Good	Mild pre-eclampsia with hydramnios.	52 hrs. Pit. Artif. rup. mem. Morphia.	High forceps at 3rd. attempt.	9/4	Fair. Convulsions on 3rd. day.	Normal.
135	IV	25	1st. of 3	Healthy. Pelvi inlet obstruction.	Med. induction at 41 wks.	26½ hrs. Hyoscine Mor. Pento.	Spont. vertex. "twilight sleep".	6/3½	Limp but responded slowly. "cerebral on 2nd. day.	Normal 1 lb. 11 oz.
150	IV	30	8th. of 10	Healthy.	Healthy but depressed. Labour induced 41 wks. Quinine and pituitrin.	5 hrs.	Spont. vertex.	7/5	Cyanosed. Marked apnoea. Convulsions 3rd. day.	Normal. 1 lb.
156	V	29	3rd. of 3	Healthy.	Post Mat. 41 wks. Mod. severe pre-eclampsia.	42½ hrs. M.distressed.	Rotation,mid cavity forceps delivery.	8/3½	Very limp. Apnoea. responded slowly.	Heavily infarcted. 1 lb. 12 oz.

In this case medical inducation of labour, on account of pelvic disproposition, was successful at the forty-first week. Labour was conducted under very heavy sedation, (twilight sleep) and after 26½ hours spontaneous vertex delivery ensued. The child, weighing 6 lbs. 3½ oz. was born cyanosed and limp, but responded to resuscitation. His behaviour was noted to be "cerebral" on the second day of life, however.

There were five cases in which both pregnancy and labour or delivery were abnormal. Vaginal haemorrhage occurred at an estimated 11 weeks gestation in Case 14, but pregnancy continued, and after four hours severe pains at term the infant was delivered by mid cavity forceps apnoeic and very shocked. He responded very slowly to resuscitation. The birth weight was 6 lbs. 12 ozs. In Case 91, pregnancy was complicated by mild preeclampsia and marked hydramnios. After unsuccessful induction with pituitrin the membranes were artificially ruptured, but in spite of the administration of morphia, pitocin and pethedine in large quantities during the 52 hour labour, severe cervical dystocia persisted. Manual rotation of the head could not be performed and eventually the cervix was incised and the large infant delivered by high forceps at the third attempt. The child's condition was considered to be fair immediately after delivery, but convulsions ensued on the second day and the child eventually showed a picture of obstructive hydrocephalus.

In Case 156 there was moderately severe preeclampsia. Delivery, at an estimated 41 weeks had to be by manual rotation and mid cavity forceps on account of prolonged labour and maternal distress. The child, who weighed 8 lbs. 3½ ozs. was apnoeic after delivery and was slow to respond to resuscitation. The placenta weighed one pound twelve ounces and was found to be heavily infarcted. In Case 87 there was mild preeclampsia and the child was delivered easily by mid cavity forceps after a labour of fourteen hours on account of poor progress. She weighed 8 lbs. 3¼ ozs. and was in good condition. The placenta is stated to have been normal.

Subsequent course of the patients.

In Table 139 is shown the progress made by the patients after delivery whether this was considered to be abnormal or not. It will be seen that four patients were apnoeic immediately after delivery, (Cases 14, 135, 150, and 156). The only other abnormal babies were those who were premature, (Cases 8 and 101), and the child who was considered to be in "fair condition" and who showed a very large cephalhaematoma (Case 91). The remaining five babies were considered to be well immediately after delivery, (Cases 27, 16, 57, 87 and 182).

Of the babies were were considered to be abnormal immediately after birth, all except one, (Case 101) in which prematurity was the only abnormality, had a previous history of abnormal pregnancy or delivery. On the other hand /

hand there was a history of abnormal pregnancy or delivery in only one of those in whom no immediate abnormality after birth was noted (Case 87).

TABLE 140

Relationship of complications of pregnancy to newborn state.

	Normal Newborn state	Abnormal Newborn state
Normal parturition	4	1
Abnormal parturition	1	6

Of the seven patients who were abnormal immediately after delivery six were noted to show other abnormalities of behaviour referable to the nervous system within one week of delivery. Convulsions occurred in three cases, (91, 150 and 156) all on the third day of life. In Case 8 the baby was noted to be drowsy, to have difficulty in feeding and to choke frequently at two days of age. In Case 14 "spasticity" was observed at two days and in Case 135 the child was "stary" and irritable on the third day. Only in Case 101 did there seem to be no abnormality until the child began to walk with support at the age of 18 months and was grossly unsteady. (Table 140)

Consideration of the birth histories and subsequent courses of patients with ataxic diplegia.

It is possible to suggest a rather frequently occurring clinical course in a proportion of patients with /

TABLE 139

Neonatal course of patients with ataxic diplegia

Case No.	Abnormalities of parturition	State after birth	1st. sign of other abnormality	Age when noted	Probable aetiological agent	Classification
8	Small antepartum haemorrhage 12 days before labour.	Premature (5 lb. 1 oz.) but good condition.	Would or could not suck at breast. Choking. Very drowsy. Slow milestones.	2 days	Unknown.	Congenital.
14	Threatened abortion. Difficult mid cavity forceps.	White asphyxia. Prolonged apnoea. (Regular respirations only after 45 mins.)	"Spastic" at 2 days. Drowsy but excitable.	2 days.	Birth injury.	Congenital.
27	None.	Well but drowsy.	Slow to hold head and sit. Slow milestones.	2 months.	Unknown.	Congenital.
91	Mild pre-eclampsia. Hydramnios. Heavy sedation. Cervical dystonia. Repeated high forceps.	Fair condition. Cephalhaematoma.	"Cerebral attacks". (convulsions)	3rd. day.	Birth injury.	Congenital.
135	Medical induction. "Twilight sleep".	Brief apnoea. Cyanosed.	"Starey" and irritable on 3rd. day of life.	3rd. day.	Birth injury.	Congenital.
150	Induced post mature labour.	Prolonged apnoea. Cyanosed. Slow to respond to resuscitation.	Restless, "cerebral", convulsions next day.	2nd. day.	Birth injury.	Congenital.
151	Moderately severe pre-eclampsia. Prolonged labour. Mid cavity forceps.	Brief apnoea. Cyanosed. "Responded slowly". Marked head moulding.	"Spasms of face and arm".	3rd. day.	Birth injury.	Congenital.
16	Varicose ulcers. Otherwise well.	Normal.	Severe unsteady. Fell frequently.	6 years.	Hereditary.	Acquired.
57	None.	Normal.	Walked unsteadily on her toes.	2-3 years.	Hereditary.	Congenital.
101	None.	Fair condition. Premature (3/8½) Required incubation.	Slow to walk and when did so on toes and very unsteadily.	18 months.	Hereditary.	Congenital.
87	Mild pre-eclampsia. Mid cavity delivery. Mid cavity forceps.	Healthy.	High pitched cry. Distended fontanelle. B.coli meningitis.	5 days.	B.coli meningitis.	Acquired.
182	None.	Healthy (10 lbs.)	Became very unsteady and clumsy after fall, and having discharging ears.	2 years.	Unknown.	Acquired.

with ataxic diplegia consisting of abnormal pregnancy and/or labour and delivery, the child being delivered apnoeic or otherwise abnormal and the proceeding to show symptoms referable to the nervous system within a few days of life. It is difficult to escape the conclusion in the light of this sequence of events, that intracranial damage may not have occurred as a result of the abnormal pregnancy and/or labour or delivery.

In the cases in which there is no such history of abnormality of pregnancy or delivery, the immediate newborn is usually normal, (the only exception being Case 101 who was not apnoeic,) and the neonatal course is not noted to be abnormal. In these patients the neurological symptoms are of later onset, for example, 6 years, 2 to 3 years, 18 months and 2 years in three of the patients. It is difficult in these patients to attribute the symptoms to birth injury, and in fact, indications of other aetiological factors are present in the majority. Thus, in Cases 16, 57 and 101 there was a family history suggestive of the hereditary transmission of ataxid diplegia. In Case 87, in which the immediate neonatal state had appeared to be normal in spite of complicated pregnancy, cerebral damage was the result of b.coli meningitis, and in Case 182 the child was normal until a fall and running ears at the age of 2 years.

The acceptance of this point of view immediately involves aetiological classification of ataxic diplegia, however, /

however, for the cases are really being distinguished on the basis of whether they have, or have not, a history suggestive of birth injury, this term being used in its widest sense.

Yet it is also convenient to classify patients with the condition according to whether neurological symptoms were present from the earliest days, congenital cases, or whether they were of later onset, and often attributable to acquired disease, acquired cases. It would be convenient if all the aetiological agents other than birth injury resulted in the later appearance of symptoms but this is not so. It would be very misleading, for example, to attribute the condition in Case 27 to birth injury, and still more those in Cases 57 and 101. It has to be recognised, therefore, that ataxic diplegia attributable to other causes than birth injury may be congenital or may be apparently acquired, as in many of the forms of hereditary ataxia.

An aetiological classification designed to take account of these considerations is as follows:

Congenital ataxic diplegia
- (a) Due to birth injury
- (b) Due to genetic factors.
- (c) Due to developmental anomalies.
- (d) Due to other causes.

Acquired ataxic diplegia
- (a) Due to genetic factors.
- (b) Due to acquired disease or trauma.
- (c) Due to other causes.

It is now necessary to consider the disorders causing /

causing these various forms of ataxic diplegia in rather more detail.

Congenital cases attributable to birth injury.

Six cases come into this category, (8, 14, 91, 135, 150, and 156). In all there is a history of abnormal parturition with an abnormal neonatal course which have been described, and are summarised in Table 139. An attempt is made to see if there is any consistent pattern in the forms of abnormalities encountered.

Only one of the six patients had any abnormality of early pregnancy, (Case 14) in which there was a threatened abortion and intermittent haemorrhage continued until term. Abnormalities of late pregnancy were present in four cases (8, 91, 150 and 156) and were associated with abnormalities of labour and/or delivery in two, (91 and 156). The abnormalities of pregnancy were antepartum haemorrhage in Case 8, preeclampsia in Case 91 and 156, associated with hydramnios in the former and a degree of post maturity, associated with the giving of large amounts of pituitrin in an attempt to induce labour in Case 150.

The abnormalities of labour were prolonged labour with inertia and the giving of pituitrin, morphia and pethedine injections during it in Case 91, twilight sleep in Case 135 and prolonged labour in Case 156. In two of these cases, 91 and 156, difficult forceps delivery followed the abnormal labour, and in Case 14, difficult mid cavity forceps delivery was required

after /

after arrest of the head during ~~after~~ apparently normal labour.

In two cases, (91 and 156) there was preeclampsia followed by extremely traumatic labour and delivery. In three cases there was a probability of hypoxia in late pregnancy, (Cases 8, 135 and 150), the only obvious abnormality. In Case 14 there was a possibility of hypoxia in early pregnancy and trauma, the result of a difficult mid cavity forceps delivery.

The only general conclusions that can be drawn are that the abnormalities appear to be concentrated in late pregnancy and labour, and delivery rather than in early pregnancy, and that multiple abnormalities are present in at least three of the patients. In general, the abnormalities of parturition which have been described resemble those found in cases of congenital hemiplegia.

Congenital ataxic diplegia due to other causes.

In the absence of any history of abnormality of parturition, of any family history suggestive of ataxic diplegia, and in the presence of a clear cut history of retarded motor development shortly after birth it is virtually impossible to say more than that in Case 27 the patient has congenital ataxic diplegia of unknown aetiology. It may be added that the case history was full and the relatives were co-operative, both parents and the sibling submitting to detailed neurological examination.

Congenital ataxic diplegia due to genetic factors. /

Congenital ataxic diplegia due to genetic factors.

The reasons for considering the girls in Cases 57 and 101 as probably suffering from hereditary congenital ataxic diplegia have been sufficiently presented when their family histories were discussed. In both cases the mother was affected, and in Case 57 the grandmother and probably the greatgrandmother as well as siblings of the mother and the patient. In both cases the suggestion is of a Mendelian dominant mode of inheritance.

In the light of the fact that the mother's brother was affected in Case 57 there seems to be no sex linkage in this case, but there is not enough information from which to draw conclusions in Case 101.

It should perhaps be stressed that the symptoms of both the patients had always shown atendency to improvement rather than worsening, and that progressive ataxia due to hereditary degenerative spinocerebellar disease seems unlikely.

Acquired ataxic diplegia due to genetic causes.

Initially it was not proposed to include the girl, (Case 16) in the series of cases of cerebral palsy. The history was of becoming ataxic at the age of about six years and progressing slowly until the age of twelve, together with the findings of spastic paraplegia and cerebellar type ataxia, with relatively little hand involvement and classical bilateral pes cavus, which the mother also showed. It was therefore /

therefore thought that she was an example of progressive spinocerebellar disease, and as she suffered from a progressive disease this should be excluded from the category of cerebral palsy. However, further observation, now for a period of five years has failed to show any progression in her symptoms or changes in her signs and she is, therefore, included as a case of ataxic diplegia, apparently of acquired type, but not certainly the result of hereditary factors.

Acquired ataxic diplegia due to acquired disease or trauma.

In case 87, the child suffered from B.coli meningitis in the neonatal period.

She was the first born of healthy parents. The mother suffered from pre-eclamptic toxaemia during her pregnancy and was admitted to hospital a few days before the delivery on this account. Labour was at term and lasted I4 hours at which time a mid cavity forceps delivery was performed, of a girl weighing 8 pounds, 3½ ounces in good condition. Five days after delivery the baby had several cyanotic attacks and developed a high pitched cry and fever (T to I00°F). B.coli was isolated from the the blood, urine and cerebrospinal fluid but treatment with Aureomycin, Chloramphenicol and Streptomycin was unsuccessful in controlling the infection. In spite of these drugs in varying combination the fever persisted, the child lost weight and failed to feed and she became progressively inadctive and disinterested, developing increasing head retraction. At the age of two months treatment with large doses of chloramphenicol systemically and into the ventricle was begun and over a period of four weeks temperature settled, the cerebro spinal fluid returned to normal and the child began to gain weight.

At the age of two years she was only slightly interested in her surroundings and appeared to be deaf. She could sit for only a few seconds without support but appeared to see objects well ~~enough~~ enough to reach them with the hand though the grasp was uncertain, tremulous and clumsy. The occipito-frontal circumference was I8" and there was gross flattening of the occiput. There was a mild convergent squint but other cranial nerves were normal.

There was moderate power in all four limbs but supination was restricted bilaterally, more on the right than the left. She could not abduct the hips fully, nor could she dorsiflex the feet. There was gross incoordination with intention tremor in all limbs. There was spastic increase of tone in all limbs, mild in the left upper limb, more severe in the right upper limb and more severe in the lower limbs. Biceps, triceps, supinator, knee and ankle jerks were all brisk, slightly brisker on the right than the left.

The plantar responses were extensor and the right Hoffmann response was positive. It was impossible to test skin sensation fully but she appeared to appreciate pin prick in all four limbs.

During the examination she had two abrupt attacks in which the head dropped, the upper limbs were momentarily elevated and the trunk flexed.....classical myoclonic attacks of flexion type.

A diagnosis of severe ataxic diplegia with mental defect, impairment of hearing (possibly of cortical origin) and myoclonic epilepsy was made.

Clinical features of ataxic diplegia.

The stages in the development of ataxic diplegia.

As has been mentioned when classification was discussed, congenital ataxic diplegia differs from diplegia in the mode of its development as well as in the final clinical picture which presents. In most patients with congenital diplegia a number of stages in a definite sequence may be defined; first the hypotonic, then the dystonic, then the rigid, and finally the spastic phase. In ataxic diplegia of congenital origin neither dystonia nor rigidity are found. A stage of marked poverty of movement associated with generalised hypotonia is usually present for a period of weeks or months after delivery. But in most cases this gives rise to little concern unless the child has other symptoms referable to the nervous system, such as convulsions or difficulties in swallowing. Only when the child is found to be unable to hold up the head, or to sit with or without support at the normal time do parents become seriously worried. In many cases there is a history that the child did not sit at the normal time and when he began to reach for objects the hands were noted to be unduly tremulous ..... commonly at the age of between five and ten months. Shortly afterwards the first signs of stiffness were noted in the limbs as the stage of poverty of movement with hypotonicity gives way gradually to the state of spasticity. The increase of tone is usually most obvious in the lower limbs and the resultant stiffness of the legs is often blamed later for the child's unsuccessful attempts to walk, his tendency to fall and his unsteadiness. In the majority of cases the onset of spasticity appears /

appears to be more gradual than in either diplegia or hemiplegia and even at the age of two or three years may not be fully developed, though unsteadiness of posture, intention tremor of the limbs and occasionally nystagmus will have been marked well before the end of the first year in most patients. Speech disturbances, most frequently of dysarthric type are rarely severe enough for the parents to complain about them until the child is over two years old, by which time most patients can stand, if only with support.

Further classification of cases of diplegia with ataxia.
(7 males and 5 females)

All the cases with diplegia and ataxia showed affection of one or both upper limbs, 9 being triplegic and 3 being tetraplegic. Cases were further classified according to the severity of their functional motor impairment which was the result of the ataxia and the paresis in association. The effect of the ataxia was to render the degree of functional impairment due to paresis of the limbs much more severe than it would otherwise have been. In slight cases the effect was merely to make the gait and the child's handling somewhat clumsy. In moderately severe cases the performance of some normal activities was made very difficult, and in severe cases activities were limited, and those that were possible were grossly clumsy. The classification of ptients with diplegia and ataxia into mildly affected, moderately severely affected and severely affected is shown in Table 141.

Five /

Table 141.

The severity of the impairment of voluntary movement and the extent of the paresis in I2 patients suffering from ataxic diplegia.

Extent of Paresis.	Mild.	Mod.Severe.	Severe.	Total.
Triplegia.	3	4	2	9
Paraplegia.	I	0	2	3
Totals.	4	4	4	I2.

Five of the patients with diplegia and cerebellar ataxia were females and seven were males. The physical findings in the twelve cases of ataxic diplegia will now be considered.

The paresis.

The paresis was triplegic in distribution in nine cases and tetraplegic in three. The severity of the paresis was not parelleled by the severity of the ataxia. The paresis was more marked in the legs than the arms in all cases. In all, voluntary movements of the limbs were more impaired in the peripheral than the proximal parts of the limbs. In particular, fine finger and toe movements were limited and weak. Because of the combination of cerebellar involvement with spastic paresis, rapid movements of the fingers and movements requiring much muscular co-ordination were poorer than would be expected in diplegic cases not complicated by ataxia.

As in hemiplegic paresis, the movements which tended to suffer most in the upper limbs were thumb and finger extension and abduction, wrist extension and supination of the forearm. In the legs, movements of the toes, dorsiflexion of the foot were the most severely affected. In severe cases weakness or limitation of extension of the elbows and knees was evident and abduction of the shoulders and hips was also affected.

As has been noted, rigidity due to contracture, was a feature of diplegia with ataxia, though rigidity due to abnormalities /

abnormalities of muscle tone itself was not. The increase of tone evident in cases showing ataxia was very variable in degree, in different cases. In those in which the paresis was slight and ataxia relatively severe, hypotonia was evident. In those with marked paresis, spastic increase of tone was evident. In two cases ankle clonus was associated with the typical pendular knee jerks of cerebellar disease. Because of the hypotonic element in the cases of diplegia with ataxia the tendon jerks showed very marked exaggeration.

Contracture of the limbs was much less marked than in cases of diplegia which did not show ataxia. When it occurred it was invariably in flexor positions, and of "hemiplegic" type.

In all the patients with diplegia and ataxia the plantar responses were extensor bilaterally.

The ataxia.

The ataxia varied in severity from mild to severe in different cases. In two cases it was predominantly right sided, in six predominantly left sided, and in four it was apparently equal in severity bilaterally.

In severe cases inco-ordination was so severe that handling objects was grossly impaired, writing was impossible and small movements very difficult. Of the eight moderately severe and severe cases of ataxic diplegia, none was able to do his own buttons or laces, and in some of these the paresis was /

Case 57. Female born 1939.

Hereditary ataxic diplegia with spastic rather than ataxic gait. Maternal grandmother, mother and sister of patient mentally retarded, suffering from ataxic diplegia and strabismus. Other relatives probably also suffer from ataxic diplegia including two or more of mother's siblings. Father alcoholic, deserted family. Mother been imprisoned on more than one occasion for soliciting. Patient had been brought up by grandmother.

Patient the result of the fourth of four pregnancies. First ended in stillbirth, and second in birth of girl with milder ataxid diplegia than patient. Third child died from tuberculous meningitis in hospital aged 4 months. Patient born at term after uncomplicated labour of five hours and spontaneous delivery. Birth weight 6 lbs. 3 ozs. Cried at once. Bottle fed and thrived. No details of motor milestones obtained. Abnormal gait only noted when she was about $3\frac{1}{2}$ years of age. "She was like the others so I thought nothing of it" said the grandmother. Attended school for mentally retarded and had made poor progress. School Medical Officer had observed child's frequent falling and gross unsteadiness, clumsiness and retarded speech development. On examination - a tall adolescent, poorly dressed dull girl, with rounded shoulders and clumsy movements.

Mentally very dull. Slow to respond to even simplest commands and questions. Speech grossly defective with many defective consonants. Slight internal strabismus, abduction of left eye being defective. Slight left facial weakness of upper motor neurone type. No asymmetry or local wasting of the limbs. Tone is slightly increased and spastic in type on the left upper limb and more increased in both lower limbs. Some hyperextensibility of fingers and toes bilaterally. Power is fair, only in the right upper limbs, definitely reduced in all movements of the left upper limbs and markedly reduced in both lower limbs. All movements are slow and accompanied by intention tremor, worse in the left upper limb and in the lower limbs than in the right upper limb. This is well produced by finger nose and heel knee testing. Whenever she moves upper limbs, associated movements of flexion or extension are seen in the contralateral upper limb. These movements are also seen when she walks Biceps, triceps, supinator, jerks are increased, brisker on the left than the right. Knee and ankle jerks more increased than the upper limb jerks. No ankle clonus. Plantar responses extensor. Abdominal reflexes equal and symmetrical. When she walks the legs are a little wider apart than they should be but the most striking features are her swaying from side to side as she proceeds and the way in which the legs swing inwards as they move forwards in circumrotatory arcs, the toes being dragged along the ground for much of the distance, though the heel touches the ground when she puts her weight on either foot. As she proceeds she elevates the arms from the shoulder level. The gait is predominantly spastic in type rather /

rather than ataxic, though there is certainly an ataxic element. When she is blindfold she is still more unsteady and deviates to the left.

No abnormalities of skin, joint, or vibration sensation could be determined.

Case 27. Male born 1945

Moderately severe ataxic diplegia with broad based stamping gait. He was the result of the third pregnancy of a mother aged thirty-one who had had one previous healthy child and one miscarriage. Pregnancy, labour and delivery were uncomplicated. He was born in hospital at term. His birth weight was 9 pounds 2 ounces. The neonatal course was uncomplicated except that it was two weeks before he regained his birth weight and he was kept in hospital until then.

He was a quiet rather inactive baby, but caused no concern until his parents became aware that his development was slower than his older brother's. He sat with support at 10 months and stood with support at the age of 18 months, but he was very slow to walk without support and seemed very unsteady on his feet. His mother wondered why his muscles were "so flabby and collapsible". He finally walked unsteadily without support shortly after the age of two years, falling a great deal, usually forwards or towards the left side. He fell less after the age of about three years.

He reached with the right hand at the age of about six months, but was jerky when he did so. He did not use the left hand until the age of two years and was more clumsy with that.

He said his first words at the age of two, but was slow to develop connected intelligible speech and even at the age of five his teacher could not understand all he said. Though his intelligence quotient was 105 on the Terman Merrill scale, on the performance scale of the Sechsler it was 67, and his school progress had been disappointing. Reading and writing were very slow to develop and he reversed small words and letters frequently. His handling remained very clumsy.

On examination at the age of eight he seemed alert and intelligent without obvious speech defect. There was a slight left facial paresis of upper motor neurone type. There was no asymmetry or wasting of the limbs. Power in the upper limbs was fair, less good on the left than the right, and less good distally than proximally. There was more marked weakness bilaterally in the lower limbs, dorsiflexion of the feet being very weak and toe movements limited. Co-ordination was grossly impaired in all limbs, much less in the right than in the left upper limb and in the upper limbs than the lower. Finger nose, and heel knee tests showed a marked intention tremor and fine and rapid movements were poorly performed. There was a constant tendency for him to over-reach with the left hand when grasping for objects. He could not do buttons or laces.

The biceps, triceps, supinator knee and ankle jerks were all /

all brisker than normal, being more exaggerated in the left upper limb than the right, and in the lower limbs than the upper. There was no ankle clonus. The abdominal reflexes were present. The plantar responses were extensor.

His gait was grossly abnormal. The legs were held wide apart and the feet were stamped to the ground as he proceeded, the heels and balls of the hallux being put to the ground simultaneously. There was considerable swaying of the body from side to side as he proceeded. He was extremely uncertain on stairs, especially when descending, and held tightly to the left banister. When blindfold, or asked to walk backwards, he placed his legs still more widely apart and deviated slightly, but consistently to the left and lowered his left shoulder when doing so. No abnormalities of superficial sensation could be demonstrated.

was relatively slight. The ataxia was most evident clinically in the broad based stamping gait of the child and in the tendency to passpoint on testing. In those with asymmetrical ataxia, the tendency to deviate to one side on blindfold or backwards walking was marked. The Romberg test was found to be consistently positive only in cases with severe ataxia. In others the compensation for the cerebellar deficiency was good enough for it to be normal. The finger nose and finger to finger tests, together with backwards walking with the eyes shut were found to be far more reliable ways of detecting ataxia.

The gait was affected somewhat differently in some cases than in others. In some the gait was on a very broad base and was stamping in type. (Case 27) In other ptients spasticity was more marked and the gait more typical of that found in diplegia as in Case 57.

In most cases a history of many falls in the first two or three years after learning to walk was elicited. Frequently the child had found going down stairs impossible for several years after he had learnt to go up, and three patients still crawled down stairs back to front though over five years old. In less affected patients the side of the banister the child held when he came down stairs often indicated the side of the ataxia. All the cases with ataxia of any severity showed much less confidence in the dark than normal children. This was presumably due to the fact that their means of visual compensation /

Case 16. Girl born 1938.

Acquired ataxic diplegia with mental impairment, probably hereditary in origin. The result of the fourth pregnancy to a mother aged 39 with marked bilateral pes cavus and slight peroneal wasting and her healthy husband. One older brother and two older sisters healthy (but not examined). Mother thinks her own mother and one of her sisters had feet similar to hers.

Patient delivered after uncomplicated pregnancy, labour and delivery. Birth weight 7 pounds. Cried at once and had entirely normal development in infancy, walking before 12 months of age and speaking at the same time.

At the age of seven she seemed to be stumbling a little when running and to be unsteady. She began to fall increasingly frequently in subsequent months, and her writing and drawing deteriorated. She had learned to do laces, but appeared to lose the knack of this. Her speech became slightly slurred, "coming in runs". She seemed to be generally less alert and interested in her surroundings and gradually became irritable and suffered temper tantrums when frustrated at school or at home. Her school work, previously good, deteriorated to such an extent that she was transferred to a school for the physically handicapped at aged ten.

She was examined in hospital at the age of 8 years and subsequently on a number of occasions. Findings were similar on these occasions to those below. She was fitted with supporting calipers in 1950, but continued to fall, though less frequently in spite of this. Her educational progress at the school for the physically handicapped was very poor, but no deterioration in her physical condition was reported by school, hospitals or her parents since about 1950.

On examination she was an obese adolescent girl with a spastic gait.

She was excitable and restless with a poor attention span and very variable obedience to commands. Her speech was scanning and slightly dysrhythmic. Both optic discs were pale. She read J4 and J4 corrected. There was nystagmus on lateral gaze.

There was $\frac{3}{4}$" shortening in the right leg compared to the left and $1\frac{1}{4}$ shortening in the right upper limbs compared to the left. The peronei were slightly wasted bilaterally. There was gross cavus and equino varus deformity of both feet. There was a slight increase of tone in the upper limbs of spastic type and a much more marked spastic increase of tone in the lower limbs. There was hypersensibility of fingers and toes. Power was slightly less than expected in the upper limbs, especially supination of the forearms, extension of /

of the digits, abduction of the digits and extension of the wrists. It was much more reduced in the lower limbs, especially distally. She could not dorsiflex the feet to within 20° of the right angle and could not waggle her toes. Eversion of the feet was very limited and weak, bilaterally more on the right than the left. Movements of the knee and hip were full in range and only slightly reduced in power.

There was intention tremor on all voluntary movements of the limbs, especially of the left upper limb, and whilst the right finger nose test could be performed with some hesitation and jerking, the left required several attempts. The heel knee tests were poorly performed bilaterally. Rapid repeated movements were poorly performed with hands and feet. She pass pointed to the left when blindfold.

Biceps, triceps, supinator jerks were much increased, being brisker on the left than the right. Knee and ankle jerks were grossly exaggerated and there was clonus bilaterally. The abdominal reflexes were absent. The plantar responses were extensor, the Hoffman signs were present.

She stood unsteadily. The Romberg test was positive to the left. She walked on her toes, with the feet in equino varus position, swinging the lower limbs, semiflexed at hips and knees from the hips and circumducting them. She dipped to the right and swayed considerably as she proceeded slowly and with difficulty, showing gross unsteadiness. Her limbs flailed about and were raised almost to shoulder level, acting as balancing organs as she walked.

No abnormalities of skin sensation to light touch, pin prick or hot and cold could be detected. Joint sense and vibration sense were (surprisingly) present.

compensation for their ataxic disorder was rendered impotent.

Other findings in the limbs.

Apparent shortening in the limbs was not a feature of ataxic diplegia. Unfortunately the number of cases is too small to confirm the impression that generalised dwarfing does not occur, but four cases were over the age of 12 years and were within normal limits for height.

The feet tended to be colder than the hands in cases of diplegia with ataxia, as in diplegia. The marked vasomotor changes found in hemiplegia were not found, however.

No disturbances of superficial sensation or in joint or vibration sense were discovered in any of the patients with diplegia and cerebellar ataxia.

Intellectual impairment.

Five of the cases were within the normal range of intelligence and two were considered ineducable. That the combination of spastic diplegia and ataxia was a particularly severe handicap from the educational point of view is indicated by the finding that only two of the five children with normal intelligence were in normal schools. This is shown in Tables 142 and 143 .

Unfortunately the numbers of cases in the survey are too small for a comparison to be made between the intelligence and schooling of cases showing ataxia alone, diplegia alone and the combination of diplegia and cerebellar ataxia.

Speech disorder.

The /

TABLE 142.

The intelligence quotients of 12 cases of diplegia with ataxia.

	115+	100-114	85-99	70-84	55-69	Under 55	Untested
Severity of paresis.							
Mild	0	0	0	1	2	1	0
Moderately severe.	0	2	1	0	0	1	0
Severe.	0	0	2	0	0	0	2
Extent of paresis.							
Triplegic.	0	2	3	1	1	2	3
Tetraplegic.	0	0	1	0	1	0	1
Totals	0	2	3	1	1	2	3

TABLE 143

School of 12 patients with diplegia and ataxia

School	Triplegic cases	Tetraplegic cases	All cases
Normal school.	2	0	2
Schools for physically handicapped.	3	1	4
Schools for the mentally handicapped.	1	1	2
Ineducable.	2	0	2
Not yet at school.	1	1	2
Totals	9	3	12

Table I45.

Speech defects in I2 patients with ataxic diplegia. Type and severity.

Extent of paresis.	No. of cases	Mild.	Mod. severe.	Severe.	No intelligible sp.	Scanning.	Total, with defects.
Triplegia	9	3'	I	2'	2	2''	8
Tetraplegia.	3	0	I	0	0	I	2
TOTALS	I2	3	2	2	2	3	I0.

' marks patients with articulatory defects and scanning speech.

The numbers of cases showing various types of speech disturbance are shown in Table 145. It will be seen that of the 12 patients suffered from speech abnormality sufficient to lead to some difficulty in comprehending what they said. Seven patients showed abnormalities of articulation. Two of these patients also showed scanning speech. The severity of the articulatory abnormalities varied from severe in which the speech was found difficult to understand even by relatives, to slight where relatively little difficulty in comprehension was encountered. One patient showed marked scanning of speech as an isolated speech defect. No cases of stutter or marked hesitation in speech were encountered.

Visual defect.

One patient showed very pale, clearly demarcated optic discs on retinal examination. The appearance suggested optic atrophy. In spite of the absence of apparent refractive error, his visual acuity was only J4 bilaterally. Two patients had very severe myopia. No cases showing amaurosis or field defect to confrontation were encountered.

Other cranial nerves. ~~(Table ——)~~.

Strabismus. Three patients showed strabismus. Two showed bilateral abducent paresis, in one complicated by unilateral third nerve weakness and impaired ability to close the eyes voluntarily. One case showed a unilateral abducent paresis. No cases showing clinically recognisable supra-nuclear paresis were found.

Facial Paresis. /

Facial Paresis. Five patients showed facial paresis of upper motor neurone type. Two of these were tetraplegic and three were triplegic. Two were considered to be cases with mild affection of the limbs, one with moderately severe affection, and two were severely affected. One case, suffering from severe triplegic paresis with cerebellar ataxia showed a very interesting bilateral upper motor neurone facial paresis. The same variations in the manifestations of the paresis found in hemiplegic and diplegic patients at rest and on emotional and voluntary movements were also encountered in cases of diplegia with ataxia.

Swallowing and tongue involvement.

One patient, ~~Case~~ referred to above, showed evidence of difficulty in swallowing. Swallowing was infrequent and drooling excessive. The same boy also showed limitation of voluntary movements of the tongue.

Epilepsy.

Three patients showed epileptic phenomena. Two of the patients were triplegic and one tetraplegic, Table 144. One case was slightly, one moderately severely and one severely affected by diplegia and cerebellar ataxia. None showed grand mal fits. One showed Jacksonian attacks and petit mal. One had salaam attacks with loss of consciousness for about 15 seconds in the attacks which occurred once or twice every day. The third case showed myoclonic jerks only.

The /

Table 144.

Epilepsy in I2 patients with ataxic diplegia.

Type of Epilepsy.		Grand Mal or Jacksonian	Petit mal.	Myoclonic Attacks.	Totals.
Extent of Paresis.	Tripleg.	I'	I'	I	2
	Tetrapleg.	0	I''	I''	I

Key ' Jacksonian attacks and petit mal.
'' Petit mal and myoclonic jerks.

The myoclonic jerks started at the age of 5 months in one case and between the ages of one year and three years in the other.

Behaviour.

One case showed hyperactive behaviour similar to that noted in hemiplegic patients and described in the section discussing the findings in hemiplegia.

Other findings.

One case showed congenital hydrocephalus, probably the result of birth injury followed by haemorrhage which had been treated by third ventriculostomy and appeared to be controlled. The head circumference when the child was examined at the age of 4 years was 24.6 inches.

One case showed scattered areas of brown skin pegmentation over the back and chest. The patches were ill-defined and irregular in shape. Their size varied from a few millimetres in diameter to 4 by 5 inches.

Family History of patients with ataxia.

The family histories of psychiatric, neurological or mental disorders are briefly indicated in Table 145. It will be seen that in the majority the parents, siblings and other relatives were healthy.

In cases 49 and 82 there was a family history of epilepsy, the father being affected in both instances, together with a sibling in the former. In Case 44 a cousin of the father was known to be epileptic, and in Case 72 the mother's brother, who may also have been the child's father, was epipeptic and psychotic, so that much of his time was spent in prisons, mental hospitals and in neurological wards. The mother was psychopathic.

In Case 148 the mother was said to be psychotic by the mental hospital which was treating her, but the impression she gave was more of agitated depression.

Siblings were abnormal in two cases. In Case 49 an elder prematurely born brother was noted to have a wry neck and a short arm on one side after delivery, but he was overseas at the time of examination and no further details or examination could be made. Another brother had recurrent afebrile convulsions as has been noted. In Case 28 an elder brother had a congenital malformation of the spine, probably a spina bifida with meningocele. It is perhaps worth noting that in Case 149 the mother had suffered from meningitis in childhood /

childhood leaving her slightly deaf, a maternal aunt had died of the same complaint and one sibling had died at the age of five from meningitis. The mother was not a carrier of meningococci or pneumococci so far as could be determined.

In Case 64 a sister of the mother had died from a congenital heart lesion in early adult life. In Case 82, two of the mother's cousins were said to have been very late in walking and to have been able to go up and down stairs only on all fours until the age of 5 or 6. Both continued to be unsteady in adult life. But it proved impossible to obtain any more reliable information about them.

Consideration of the family history.

In six of the fifteen cases there was no suspicion of any abnormal congenital conditions in the family. In Case 148 the mother was psychotic. In Case 166 no family history could be elicited. In Case 64 the relative with a congenital heart is probably coincidental. The dead siblings were the result of premature delivery and infections in the first year. In the remaining six cases the family history is probably more relevant. In Case 149 the apparent family predisposition to develop meningitis may have been of aetiological importance in the patient developing meningitis and becoming ataxic as a result. In Cases 28, 44, 49, 72 and 82 there was a history suggesting congenital abnormalities of the nervous system. This resulted in epilepsy in three cases, whilst in Case 28 an elder brother had a definite malformation

of the spine and in Case 82 there seems no doubt that two cousins of the mother had cerebral palsy of some description. That as many as a third of the patients should have a history suggestive of congenital neurological abnormalities suggests, at least, that there may be some underlying predisposition to develop neurological abnormalities in the families of ataxic patients, which the patients themselves may well share. On the other hand, with the possible, but unlikely exception of Case 82 it is impossible to demonstrate that there is any tendency for other members of the family to be ataxic.

TABLE 145

Social and Family History of Ataxic Patients

Case No.	Social Class.	Preg. No.	Employment Father	Employment Mother	Broken Home	Antenatal care.	Place of birth	Father	Mother	Siblings	Relatives
28	V.	2/3	Intermittent F. minded	Part-time Unintelligent	No	Regular	Hospital B.	Chronic Nephritis	Healthy	1 healthy	Healthy
30	III	1/1	-	Unknown	Illegit. Adopted.	Unknown	Hostel B.	Unknown	Healthy	-	Unknown
33	IV	1/1	Regular	No	No	Regular	Hospital B.	Healthy	Healthy	-	Healthy
44	III	1/3	Regular	No	No	Regular	Hospital B.	Healthy	Healthy	2 healthy	F.epileptic
49	III	6/6	Regular	No	No	Regular	Home	Child-hood conv.	Healthy	1 wry neck short arm	Healthy
64	IV	6/8	Regular	Part-time	Parents separated	Regular	Home	Healthy	Healthy	6 dead 1 healthy	Aunt with cong.heart
73	IV	1/1	Unknown	Whole-time	Illegit.	Irregular	Home	Unknown	Healthy	-	Healthy
72	VI	1/2	Unknown	Part-time	Illegit.	Irregular	Hostel	Unknown	Psycho-pathic.	1 healthy	Healthy
78	II	2/2	Regular	No	No	Regular	Home	Healthy	Healthy	1 healthy	Healthy
82	IV	1/3	Regular	No	No	Regular	Hospital B.	Epileptic	Healthy	2 healthy	2 M's cousins abnormal.
115	III	1/1	Regular	No	No	Regular	Hospital B.	Healthy	Healthy	1 healthy	Healthy
148	III	1/2	Regular	No	No	Regular	Home	Healthy	Psychotic	-	Healthy
149	III	2/4	Regular	No	No	Regular	Hospital B.	Healthy	Healthy	1 dead 2 healthy	Aunt died of meningitis.
179	III	2/3	Regular	No	No	Regular	Home	Healthy	Healthy	2 healthy	Healthy

B. = booked

History of the other pregnancies of the mothers with children suffering from ataxia.

There was a total of 39 pregnancies to the 15 mothers of patients with ataxia, giving a fertility of 2.6 per mother, which is approximately the same as that for congenital hemiplegia, diplegia and dyskinesia. It is lower than that for acquired hemiplegia and ataxic diplegia.

Of the 24 pregnancies other than those resulting in the birth of the patients, two resulted in miscarriages (Cases 49 and 148). Six were complicated, by prematurity in four instances in Case 64, resulting in the neonatal deaths of all four babies, by premature breech delivery in Case 49 and by pyelitis in Case 179. Sixteen, or two-thirds of the 24 were normal. (Table 147)

No stillbirths occurred, but four very premature infants died in Case 64 within a few hours of delivery, and there were also two postneonatal deaths to this mother, one from bronchopneumonia, and one from gastroenteritis. Three siblings were congenitally abnormal, one with an abnormal back in Case 28, one with wry neck and a short arm, and one epileptic in Case 44. There was a total of 15 healthy siblings from the total of 24 other pregnancies or approximately 58%, a result comparable to that for ataxic diplegia and congenital hemiplegia.

Spacing of pregnancies.

This did not follow the pattern seen in the other forms of cerebral /

cerebral palsy though the number of pregnancies is too small for conclusions to be drawn from this.

	Time	Number of cases
Average lapse of time between previous deliveries.	2.1	7
Between last prior delivery and and birth of patient.	1.7	7
Between birth of patient and subsequent delivery.	2.7	7

It will be seen that the lapse of time between the birth of the last child and the delivery of the patient was in fact shorter than the lapse of time after the patient and the next pregnancy and between prior deliveries. No very obvious reason for these findings can be suggested.

Number of pregnancy resulting in birth of patients.

In Table 146 is shown the distribution of the patients by the number of the pregnancy resulting in their birth.

TABLE 146

	Observed	Approx. Expected
1st. born and only.	4	6
1st. born of number.	4	
2nd. born	4	4
3rd. born	0	3
4th. born	0	1
5th. born	1	.5
6th. born	1	
(last born)	6	

It will be seen that more than half the patients were first born but the distirubition is now significantly different /

TABLE 147.

The other pregnancies of mothers of ataxic patients

Case No.	Social Class	Age of Mother	No. of Preg.	Misc.	Other abnormal parturition	Often normal	Stillbirth	Neonatal deaths	Post neonatal deaths	Abnormal siblings	Normal siblings
28	V.	26	2/3	0	0	0	0	0	0	1 abnormal back	1
30	III.	?	1/1	0	0	0	0	0	0	0	0
33	IV.	39	1/1	0	0	0	0	0	0	0	0
44	III.	21	1/3	0	0	2	0	0	0	0	2
49	III.	40	6/6	1	1 prem.breech	2	0	0	0	1 wry neck, short arm. 1 Epileptic	2
64	IV.	24	6/8	0	4 premature	4	0	4	2	0	1
73	IV.	23	1/1	0	0	0	0	0	0	0	0
72	III.	17	1/2	0	0	1	0	0	0	0	1
78	II.	28	2/2	0	0	1	0	0	0	0	1
82	IV.	27	1/3	0	0	2	0	0	0	0	2
115	III.	31	1/1	0	0	0	0	0	0	0	0
148	III.	25	1/2	1	0	0	0	0	0	0	0
149	III.	26	2/4	0	0	2	0	0	0	0	1 died meningitis. 2 healthy.
179	III.	25	2/3	0	1 pyelitis	1	0	0	0	0	2 healthy.

different from that expected on the basis of chance in the group.

CHAPTER 9b.

Birth histories of patients with ataxia.

The birth histories of the 15 ataxic patients are summarised in Tables 148,149. The cases were divided into those in which the ataxia appeared to date from birth, numbering nine, and those in which it appeared to be acquired, usually as the result of disease, in later childhood and in which the children were thought to have been normal for a period after delivery, numbering ~~five.~~ six. Unfortunately the pregnancies and deliveries of the patients with acquired ataxia are less well documented than those of the congenital group. However, it seems that apart from premature rupture of membranes in Case 64 and forceps delivery in Case 115, the parturition was normal in these patients. Only one of them, Case 115 appeared to be abnormal immediately after birth. Although he cried at once the child did show considerable bruising of the head and prominent forceps marks. The neonatal course was stated to be normal in all cases.

In the congenital group there is a much higher incidence of abnormality of parturition, only one of the 9 patients, Case 78 having apparently normal pregnancy and delivery. The child breathed at once after birth, though hydrocephalus was apparent from the start.

On the remaining 8 cases, all except Case 73 had abnormalities of pregnancy. Pre-eclampsia was present in three /

TABLE 148a

The birth histories of patients thought to be suffering from congenital ataxia

Case No.	Mat. Age	No. of Preg.	Maternal health	Pregnancy	Labour	Delivery	Bth. Wgt. lbs.	Bth. Wgt. oz.	Neonatal condition	Placenta
28	26	2	Good.	Mod. sev. pre-eclampsia.	At 32-33 wks. 6½ hours.	Spont. vertex.	5	2¾	Cried at once. "Rather feeble initially".	Healthy.
33	39	1	Ovarian cyst removed 6 wks.	Mild pre-eclampsia.	At 38 wks. 1¼ hrs. Foetal distress.	Low mid cavity forceps	5	8	Brief apnoea but colour good. Cyanotic attacks next day.	Much infarcts. 1 lb. 7 oz.
49	40	5	Healthy.	Antepartum haem. 7 mths. Mod. sev. pre-eclampsia.	30 hours.	Spont. vertex.	7	8	Good.	Normal.
73	23	1	Healthy.	Normal.	48 hours.	Difficult forceps.	70	-	Normal except for large head. Cyanotic attacks from age 3 days.	Unknown.
78	28	2	Healthy.	Normal.	"Normal"	Spont. vertex.	Mature		Normal except for hydrocephalus. Fits aged 8 weeks.	Unknown.
82	27	1	Healthy.	Fainting 3-4 times a day from 4 mths. onward. Antepartum haem at 6½ mths.	Prolonged 1st. stage 32 hrs. 2nd. stage 20 min.	Spont. vertex.	7	5	Cyanosed. Petechial rash Jaundice within 24 hrs.	Normal.
148	25	1	Psychotic but otherwise well.	Attem. version 28 wks. Another at 36 wks. followed by onset labour.	10 hours.	Breech extraction.	4	4	Cried at once.	Unknown.
179	25	2	Pyelitis.	Threatened abort. 2 mths. Pyelitis. False labour near term.	Induced at term 24 hrs.	Spont. vertex.	6	8	Prolonged apnoea. Resp. difficulty. Very quiet.	Unknown.
44	21	1	Good but contracted pelvic outlet.	Mild pre-eclamptic toxaemia.	Prolonged 2nd. stage.	Diff. mid-cavity forceps deliv.	8	14	Prolonged apnoea. Marked caput. Cyanotic attacks next day.	Normal. 1 lb. 7 oz.

TABLE 148 b.

Immediate neonatal condition and neonatal course of the patients with congenital ataxia

Case No.	Abnormality of parturition.	Immediate neonatal state.	Bth. Wgt. lbs.	oz.	Subsequent course.	Subsequent progress.
28	Moderately severe pre-eclampsia. Prem. onset of labour.	Cried at once.	5	2¾	Noted to be rather feeble for first few days. Fed poorly.	Floppy baby. Retarded milestones.
33	Ovarian cyst removed at 5 wks. Mild pre-eclampsia. Foetal distress during labour. Low mid cavity forceps.	"Shocked".	5	8	Poor feeding. Irritable and restless.	Very regarded milestones.
49	Mod. severe pre-eclampsia. Antepartum haem. 7 mths. Labour 30 hours.	Good.	7	8	Very quiet. Sleepy. Poor feeder.	Continued to be very sleepy. Retarded milestones.
44	Mild pre-eclampsia. Prolonged 2nd. stage. Diff. mid cavity forceps.	Prolonged apnoea. Marked caput.	8	14	Twitching attacks and cyanosis from second day for 1 week.	Floppy hypotonic baby with retarded milestones.
73	Prolonged labour. Difficult forceps.	Breathed at once. Large baby. Hydrocephalus.	10	0	Cyanotic attacks at 3 days and 7 days.	Increasing size of head. Hypotonic. Retarded milestones.
78	Normal.	Good but hydrocephalus.	"Mature"		Normal except for increasing head size.	Increasing size of head. Convulsions from age 8 wks. Regarded milestones.
82	Fainting 3-4 times a day from 4 mths. gest. Antepartum haem. at 6½ mths. Prolonged 1st. stage. Precipitate 2nd. stage.	Cyanosed but breathed at once.	7	5	Petechial rash and jaundice within 24 hrs.	Retarded milestones.
148	Attempted version 28 wks. and again at 36 wks. followed by prem. onset of labour. Breech extraction.	Cried at once. Premature.	4	4	Slight jaundice. Normal progress.	Hypotonic, placid baby. Retarded milestones.
179	Threatened abortion at 2 mths. gest. Induced at term. 2nd. stage 24 hours.	Prolonged apnoea. Very cyanosed.	6	8	Persistent respiratory difficulty for 2 days. Irritability.	Much irritability. Retarded milestones.

three cases, 28, 33 and 49 and was complicated by antepartum haemorrhage in Case 49. There was threatened abortion at an estimated two months' gestation in Case 179, but otherwise early pregnancy was normal in all the cases. In Case 82 the mother had recurrent fainting during the whole of her pregancy after the first four months, and in some of her attacks she was unconscious for a matter of some minutes according to witnesses. It is possible that these attacks were epileptic, but they ceased after pregnancy, and it seems more likely that they were syncopal in origin. She had a slight antepartum haemorrhage at an estimated 26 weeks' gestation. In Case 148, version at 28 weeks was thought to be successful, but was repeated with great difficulty at an estimated thirty-six weeks' gestation. Labour ensued within a few hours and a difficult breech extraction was performed. The child cried at once.

Abnormalities of pregnancy were associated with abnormalities of labour and delivery in three cases. In Case 33 there was foetal distress during labour after the pre-eclamptic pregnancy, and a rapid low mid cavity forceps delivery was performed. The child had brief apnoea, but thereafter seemed well until cyanotic attacks were apparent about 18 hours later. In Case 49, a fifth pregnancy labour lasted for thirty hours before spontaneous vertex delivery of an apparently normal infant occurred. In Case 82, fainting during /

during pregnancy and antepartum haemorrhage were followed by a very prolonged first stage of labour and a precipitated second stage lasting only 20 minutes. The child was cyanosed at the time of delivery and soon showed a generalised petechial rash and jaundice within 24 hours. In Case 148 attempted external version was followed by the onset of labour with the foetus still as a breech presentation and a breech extraction was performed of a premature child at an estimated 36 weeks gestation. In one Case (73) a difficult high forceps delivery was achieved after a labour of 48 hours. The child was noted to be hydrocephalic at birth and it seems likely that this, together with the large size of the infant, was the cause of the slow labour.

Immediate neonatal state and neonatal course of ptients with ataxia.

Of the acquired cases, all made normal progress during the neonatal period and all ~~five~~ six appeared to be normal immediately after delivery with the exception of Case 115 in which there was considerable bruising to the head which has already been briefly described. Attention, will, therefore, be concentrated on the congenital cases. Table 148b.

Six of the nine patients breathed at once after birth, (Cases 28, 49, 73, 78, 82 and 149), though in Cases 73 and 78 the children were abnormal in having hydrocephalic heads noted at once by the attendant doctor (though in Case 73 he did /

did not mention this in his admission letter to the hospital when the child was thirty months of age). In Case 82 the child was cyanosed, hypotonic and feeble for some hours after birth, though as noted he breathed at once.

In the other three cases there was apnoea after birth. This was brief in Case 33, but prolonged in Cases 44 and 179.

The subsequent behaviour of the patients in the neonatal period, because of cyanotic attacks which persisted for a week in Case 73, because of twitching attacks and "spasms" which consisted of sudden extension of the trunk and limbs in Case 44, and because of drowsiness and poor feeding in Cases 48 and 49. In Cases 82 and 148 there was drowsiness and reluctance to feed associated with jaundice which cleared within 10 days and was associated with petechiae appearing within six hours of delivery in Case 82. In Case 78, increasing head size was noted even during the neonatal period, and in Case 179 there was persistent respiratory difficulty for three days, followed by marked irritability which was considered to be characteristic of cerebral injury at the time.

After the first month, generalised hypotonia was noted in four cases, (28, 44, 148 and 73). In all these patients milestones were retarded, and in addition, the head steadily grew in size in Case 73. In Case 78 convulsions, with no loss of consciousness, consisting of alteration of colour, irregular movements of all limbs and irritability were associated /

associated with steady head enlargement. In Case 179, irritability was also present in association with retarded motor development, but in the remaining two cases, (33 and 82), retarded development was the only symptom noted by the parents at the time.

It will be observed that the abnormalities present in the late neonatal period in these patients were not really striking, and in fact only constant and sometimes direct questioning elicited some of the information which has been presented. Unless the child presented with convulsions or a steadily enlarging head, his retarded progress and poverty of movement tended to be attributed to his having "had a bad time during birth". Nevertheless, the fact that the children were described as "floppy" in four cases, all of whom showed retarded motor development, is probably significant.

The aetiology of acquired ataxia.

The histories of the patients with acquired ataxia are summarised in Table 150. It will be seen that tuberculous meningitis was responsible in two cases, pyogenic meningitis of unknown type in one case and whooping cough encephalopathy in Case 30. In Case 64 the origin of the ataxia remained obscure. In Case 115 ataxia followed neurosurgery.

Both cases of tuberculous meningitis were severe, and both had intermittent spinal block for a period of some weeks. (Case 64 during his relapse when the organisms were found to have /

TABLE 150

The aetiology of acquired ataxia

Case No.		Progress in 1st. year	Onset of illness.	Nature of illness.	Course.	Residual disability.
30	Premature	Good.	Age 5	Whooping cough. Encephalopathy after 2 weeks.	Convulsions. Coma. Fever.	Disc. from hospital after 4 mths. when thought to be mentally defective. Gradual improvement. Residual ataxia espec. to right.
64	Premature rupture of membranes. Seemed normal in neonatal period.	Good to 9 months.	age 9 mths.	Tuberculous meningitis 1950.	Treated for 6 mths. Then relapsed and had second course of treatment 1951. Streptomycin treatment.	Severe ataxia and deafness.
72	Normal.	Good.	3 yrs.	Unknown. Began with convulsions 8-10 times a day until whooping cough aged 5 years.	Became unsteady after 1 year of fits and remained so.	Moderately severe ataxia. Hyperactivity. Mental defect.
149	Normal.	Good until aged 5 wks.	5 wks.	Meningitis of unknown type.	Hospitalised after being ill for 10 days, for 6 weeks. Slow improvement.	Optic atrophy 6th, 7th. nerve paresis. Partial deafness. Bilateral ataxia.
166	Unknown.	Good.	14 mths.	Severe tuberculous meningitis.	Hospitalised after 3 wks. illness for 1 year. Streptomycin treatment for 8 months.	Severe cerebellar ataxia. Minimal left hemiplegia. Mental impairment.
115	Forceps delivery	Delayed motor milestones by 1 year stage	9-12 mths	Removal of large mid-line cerebellar tumour at 30 mths.	Gradual improvement to present level.	Severe cerebellar ataxia.

have developed partial resistance to streptomycin, and in Case 149 for a period of five weeks after she had been ill for four months. In both, intrathecal as well as systemic streptomycin was given together with oral P.A.S. In neither case was dihydrostreptomycin used.

In Case 64, the child became deaf during his relapse and caloric testing revealed that gross vestibular damage had also been sustained, though caloric reaction and hearing were normal in Case 166 in spite of the very prolonged treatment. Thus, though in the former case much of the ataxia might be attributal to vestibular loss, in the latter it was probably purely cerebellar in origin.

In Case 30 the whooping cough encephalopathy is well documented apart from the fact that the cerebro-spinal fluid results were lost.

In Case 149 the child was treated by penicillin injections and sulphonomide by mouth, but unfortunately other details of his hospital admission, apart from the fact that he is recorded as having meningitis, are missing. He was certainly very ill when sent to hospital, having been fevered and refusing feeds for ten days at the age of 5 weeks. On examination he showed severe bilateral ataxia, optic atrophy and impaired hearing though mentally he was alert and interested.

In Case 72, the history is compatible with subacute sclerosing leucoencephalitis or other similar subacute encephalitis or with cerebral thrombophlebitis, but it is impossible to make a firm retrospective diagnosis.

In Case II5 the patient was first thought to be abnormal at the age of about 9 months,because he was slow to sit without support.He finally achieved this at the age of about one year.He walked only with the support of two people at the age of two.His hands had always seemed to be clumsy.From the age of I8 months to two years he failed to make any developmental progress and after this retrogressed,losing the ability to feed himself with a spoon or even crawl.He was finally referred for neurosurgery at the age of thirty months.A posterior fossa tumour was removed which was cystic in type and involved both lobes of the cerebellum.Following operation his ataxia greadually became less marked,but he was still unable to walk without support by the age of five.He could feed himself,however and was scribbling with a pencil.

The Clinical findings in 15 cases of ataxia.

Fifteen children with cerebral palsy were classified as suffering from ataxia of the limbs as their predominant disability. In 14 of the patients the ataxia was cerebellar in type. In one, the ataxia was mixed cerebellar and vestibular in type, the result of tuberculous meningitis treated with streptomycin.

Of the fifteen cases, nine were males and six were females. The severity of their ataxia, its distribution and the clinical findings in the patients varied greatly. All the cases showed impairment of co-ordination of the limbs, however, and in one was paresis of the limbs any more than a slight disability. Power of the limbs tended to be within normal limits. Use of the power was defective.

The Early Course of Cerebellar Ataxia.

There was a great similarity in the descriptions of the behaviour of patients suffering from congenital ataxia and these with congenital ataxic diplegia. Birth injury and its immediate effects appeared to be rather more frequent amongst the ataxic patients, but the subsequent history of poverty of movement associated with generalised hypotonia, retarded motor development and slowness in holding up the head, sitting and getting on to the feet was common to both groups. The same jerkiness of movement, tremor or clumsiness which was often observed when patients with ataxic diplegia /

TABLE 153

The extent and severity of ataxia in 15 patients

Severity of ataxia	Slight	Moderately severe.	Severe	Totals
Direction of ataxia				
Predominantly right.	1	1	0	2
Predominantly left.	1	3	0	4
Bilateral.	2	4	3	9
Totals.	4	8	3	15

TABLE 154

Associated neurological abnormalities in the limbs in 15 patients with ataxia.

Severity of ataxia.	No. of patients	Increase of muscle tone: Paraplegic.	Increase of muscle tone: Hemiplegic.	Involuntary movement: Athetosis.	Involuntary movement: Tremor	Hypotonia: Unilateral	Hypotonia: Bilateral
Mild.	4	0	1	0	0	2	0
Mod. severe	8	1	4	1	2	1	1
Severe	3	1	1	0	0	1	0
Totals	15	2 °	6 ⁑	1	2	4	1

° One with bilateral Babinski signs.

⁑ Three with unilateral Babinski signs.

diplegia first tried to reach for objects was also apparent in those with cerebellar ataxia. Curiously enough it was rarely a cause of great concern to parents at the time. Usually they only brought the patient for medical care when it was apparent that he was unable to stand or walk at the age of eighteen months or more. Complaints were frequently that though the child had been able to walk with support for some months, he had been quite unable to take any steps unsupported, that his gait seemed abnormal and that he was very liable to fall. Complaints of great clumsiness in handling objects were usually incidental, though slow and indistinct speech were commonly noted with concern.

The rather longer period which elapsed before patients with ataxia were referred for treatment compared to those with ataxic diplegia may have been related to the absence in the former of the alarming finding of spasticity in the legs.

The extent of the ataxia and its severity.

The ataxia was central in type in 9 cases as shown in Table 153. In these, the child was more or less unsteady on the upright position, but tended to deviate to one side no more than to the other. The severity of the ataxia of the limbs was not necessarily quite so symmetrical and most of these cases showed somewhat greater inco-ordination of voluntary movement on one side than on the other, though all the limbs were affected to some degree.

In six patients the ataxia was predominantly unilateral, the /

the child tending to deviate and passpoint to one side more than the other. Two cases showed predominantly right-sided ataxia and four left-sided ataxia. Though the limbs on the affected side in these patients showed inco-ordination on voluntary movement, the opposite limbs also showed some impairment of co-ordination, less marked in degree.

Four patients were classified as suffering from ataxia of mild degree. In these, normal activities were possible though they might be somewhat clumsy and motor milestones retarded to some extent. Eight patients were classified as having ataxia of moderately severe degree. In these patients some normal activities were impossible and others impaired, but the ataxia was not severe enough to prevent independent gait and writing. Severe cases showed great impairment of their ability to perform normal tasks. Their gait was grossly unsteady, or they had no independent walking and their manipulations were extremely clumsy, so that their writing was extremely defective. They required help in dressing.

The manifestations of ataxia.

In the majority of cases the most evident manifestation of underlying ataxia was abnormality of the gait rather than gross unsteadiness in the erect position. The legs tended to be wide apart, the feet to be stamped to the ground and the steps to be small. In cases in which the ataxia was more marked to one side than to the other, the shoulder of the more affected side tended to be lower and more forward than /

than that on the affected. In two cases the difference in the levels of the shoulders, was as much as 2". The back showed a scoliosis in the thoracic region towards the less affected side and the head was tilted towards it, with a scoliosis in the cervical region towards the affected side.

That the appearance of steadiness which most of these children showed was the result of walking on a broad base and compensating by visual means for their ataxia could easily be demonstrated. When requested to walk along a straight line and to place their toes upon it, a marked tendency to fall was often evident. When asked to walk with the eyes closed or blindfold the majority of the cases were very unsure and tended to fall to one or other side. This was especially evident when the child was asked to walk backwards blindfolded, and ataxia was much more evident on this test than on the Romberg test, which was found to be rather unreliable. It was positive in only 5 cases, where as walking backwards blindfold revealed ataxia in all those in the series.

The majority of cases with ataxia showed delay in walking, the average age of achieving independent walking in 10 cases thought to be of congenital origin was 2 years 3 months. Three cases walked independently only after the age of 3½ years. There tended to be a period of three or four months during which the child would walk holding on to furniture, but not without /

without support. When independent walking was achieved, most children fell extremely frequently, and the knees of most cases of ataxia bear numerous scars, the result of this early marked unsteadiness. At a somewhat later stage the patients have the greatest difficulty learning to manage stairs and especially to descend them. The side of the stairs to which the child holds in his descent was found to be a good guide to the side on which the ataxia was more severe. Two cases with bilateral ataxia still descended stairs hind and foremost, on all fours at the age of six.

As when they were blindfolded, cases of ataxia tended to be uncertain when it was dark. ~~Though~~ Predominantly unilateral cases could walk remarkably straight with their eyes open and their legs wide apart. As soon as they were deprived of the use of their eyes, very marked deviations to the affected side resulted in all except those who were slightly affected. In all patients with ataxia, however, there appeared to be a very poor sense of direction, and in a school for spastics, ataxia cases may frequently be seen to go to the wrong end of corridors after three or four years of residence. Their poor sense of direction was also shown in the loss of direction shown in the writing of cases of ataxia, and its tendency to stray from the horizontal on the page.

On testing, all of those with predominal unilateral ataxia showed pass pointing towards the ataxic side. When the arms were held forwards and extended, the limb on the affected side showed /

showed falling away before the opposite arm. In bilateral cases the posture of the limbs was poorly maintained and the falling away was usually less consistently to one side or the other. Similarly, cases with predominantly unilateral ataxia tested by the finger to nose to observer's finger test showed pass pointing consistently in one direction.

Cases with bilateral ataxia showed grossly defective sense of position rather than true consistent pass pointing in any one direction. The result of ataxia in the upper limbs was that the children were unable to reach for objects without looking where they were putting the hand. This may sound a particularly severe disability in itself. But it is only necessary to watch an ataxic child drawing and rubbing out what he had drawn in the wrong direction, then look and reach for his pencil, then look and reach for his rubber to realise the time he wastes in his fruitless and abnormal activity.

Disturbances in the limbs in the patients with ataxia.

Clumsiness of movement was invariable in patients with ataxia. It affected fine and rapid movements, and therefore, movements of the distal parts of the limbs, to a greater extent than corse movements. In particular, fine and rapid movements of the fingers were impaired and of the 10 congenital cases, buttons and laces could only be managed without help in six, and in none of these before the age of five. The clumsiness /

TABLE 157

Associated neurological abnormalities in the limbs in 15 patients with ataxia

Severity of the ataxia.	Number of patients	Increase of muscle tone		Involuntary movement		Hypotonia	
		Paraplegic	Hemiplegic	Athetosis	Tremor	Unilateral	Bilateral
Mild	4	0	1	0	0	2	0
Moderately severe	8	1	4	1	2	1	1
Severe	3	1	1	0	0	1	0
Totals	15	2 °	6 ⁑	1	2	4	1

° One with bilateral Babinski signs.

⁑ Three with unilateral Babinski signs.

Case 30. Girl born 1939.

Acquired ataxia the result of whooping cough encephalopathy. The first illegitimate child of a healthy father and asthmatic mother, born before term after uncomplicated pregnancy labour and delivery. Birth weight was between 4 and 5 pounds. Bottle fed and thrived. Entirely normal development during infancy and was well until she acquired whooping cough two weeks after beginning school at the age of five. Ten days after onset she developed a very high fever, became progressively drowsy and within three or four hours was comatose, with staring eyes, unable to swallow. She was transferred to hospital where she was desperately ill for some weeks. Was tube fed for ten weeks, but then spoon feeding again became possible and she began to recognise her mother again. She was eventually returned home after 15 weeks, still unable to speak, but taking an intelligent interest in what went on around her. Her limb movements seemed weak and poorly co-ordinated, and though she could get up into the sitting position herself, she was unable to get up or down from it unaided. She had neither bowel nor bladder control. She said her first word about one year after the onset of her illness, and gradually added to her vocabulary. By that time she was able to take solids again. She continued to feel floppy and was unable to stand if put on her feet until two years after the onset of her illness. Even then her mother observed that all her movements were clumsy and jerky, though the right arm was more affected than the left.

By the age of eight she was able to feed herself again, talk intelligibly, and do much of her own dressing, but as she did more with her hands the effect of a tremor in them became more apparent, especially in writing. She has steadily improved since then, however. At her school for the physically handicapped she falls less often than she did, writes along lines, and more clearly than she did.

On examination she was a plump ataxic adolescent. She was pleasant, obedient and obviously rather dull girl with little initiative, but with a sense of humour. There was marked scanning of her speech and sometimes a definite explosive quality was apparent. There was no asymmetry or localised wasting in the limbs. There was slight hypotonus in both upper limbs, more evident on the left than the right, but the tone of the lower limbs was within normal limits. Power was within normal limits in the left upper limb, but slightly reduced, especially distally in the right. Power of finger abduction and adduction, finger and wrist extension and supination of the forearm was slightly reduced. Power in the right lower limbs was similarly slightly less good than that of the left. All voluntary movements were full in range.

Co-ordination /

Co-ordination was impaired in all four limbs, movements being jerky and accompanied by intention tremor, made very obvious by the finger nose, or heel knee tests, especially on the right. Fine repetitive movements of the fingers and toes or forearms are very poorly performed, worse on right than left. Fine movements like stitching, doing buttons or fastening a suspender are poorer on right then left.

When she stands she sways to the right. When she walks she does so on a broad base with increased body sway and the right shoulder lower than the left. There is a slight associated position of the right upper limb similar to that found in hemiplegia, adducted and flexed at the shoulder, flexed at the elbow, pronated at the forearm and flexed at the wrist and fingers.

The biceps, triceps, supinator jerks are within normal limits, but slightly brisker on the right than the left. The knee jerks are brisk and pendular. The ankle jerks are moderately brisk. The right plantar response is equivocal. The left if flexor. No abnormalities of skin sensation, joint or vibration sense were detected.

clumsiness of movement tended to be noted in congenital cases at an early age, and in seven the child was stated to have been clumsy with one or both hands before the age of one year. The clumsiness was marked only on the more affected side in the cases showing predominantly unilateral ataxia, but on examination it was usual to find that some degree of dysdiadochokinesia was also present in the fingers of the opposite side. (Table 154)

Intention tremor was one of the most consistant features of patients with ataxia, and it was most evident when the child reached for objects. In two patients with predominantly unilateral ataxia it was much more marked in the upper limb on the affected side, but was present bilaterally in all nine cases of central ataxia. It was variable in severity in different cases, but the clinical impression was that it tended to be much more severe in younger than in elder patients. In the patients in whom intention tremor was present in the upper limbs it could also be elicited in the lower and tended to be more severe there in the majority, possibly because the children had not the same experience in trying to reduce it to the minimum by voluntary motor activity.

Well marked hypotonia was present in five patients, three of them cases of predominantly unilateral ataxia and two cases of bilateral ataxia. All the cases of unilateral ataxia showed unilateral hypotonia, on the side to which the ataxia was evident. One of the cases of bilateral ataxia showed hypotonia only on one side while the other showed generalised hyptonia. /

hyptonia.

The reason why a higher proportion of cases did not show more marked hypotonia is that many had other minor neurological abnormalities of upper motor neurone type, resulting in slight spastic increase of tone replacing hypotonia. In two patients no neurological abnormality could be discovered other than ataxia, however, and tone in the limbs was normal. Two of these cases were severely and one slightly affected by ataxia. The types of upper motor neurone lesion encountered which gave rise to slight increase in the tone of the limbs were hemiplegic in type in six cases and paraplegic in two cases. These findings are shown in Table 154.

The patients showing hemiplegic increase in tone were somewhat similar in type. Apart from slightly greater impairment of fine and rapid movement of the fingers on the affected side and slight weakness of supination of the forearm, finger extension and movements of the thumb, paresis was not a striking feature. On voluntary activity of the contralateral arm, however, associated movements of the flexor types were present in the hemiplegic upper limb, the hand clenching, the forearm pronating and the elbow flexing slightly. On examination of the tone of the limbs on the hemiplegic side some increase of tone of spastic type was evident. The biceps, triceps, supinator, knee and ankle jerks were increased out of proportion to the degree of increase of tone. The knee jerks in some cases were exaggerated and seemed to show

a /

a mixture of spastic increase of power and pendular increase in range of the response to stimulation.

Three of the patients showed extensor plantar responses on the hemiplegic side. In three cases the responses were equivocal or flexor. Four patients showed cold hands and feet on the side affected by hemiplegic increase in tone.

One case showing bilateral ataxia with mild hemiplegia showed slight athetosis of the fingers on the affected side. Only one patient showed apparent shortening in the hemiplegic upper limb of more than $\frac{1}{2}$" and in her case it was $\frac{2}{3}$". Paresis of her affected arm was very slight, and the limb was useful.

Two cases showed slight paraplegic increase of tone of spastic type in each case. Both were cases of moderately severe bilateral ataxia, and both were cases of congenital hydrocephalus. In neither case was the increase of tone in the lower limbs more than slight, and since paresis of the limbs could not be demonstrated it was felt that they should be justified as cases of ataxia and not diplegia with cerebellar ataxia. One case showed extensor plantar responses bilaterally. One of the patients showed a lower temperature in the feet than the hand. Neither case showed apparent dwarfing.

Apart from the case previously referred to, no asymmetry of the limbs was evident. No cases other than those already mentioned showed differences of temperature in the limbs.

Neglect /

Neglect of the affected limbs was not a feature of ataxia.

One case of ataxia following tuberculour meningitis showed partial superficial hemianaestheia to all modalities and impairment of joint and position sense. The patient showed predominantly unilateral ataxia of moderate severity associated with slight hemiplegic increase of tone, on the same side as that on which the sensory impairment was present.

Intellectual impairment in ataxia.

The intellectual impairment in the cases of ataxia as measured by different tests given in different circumstances by different testers are shown on tables and it will be observed that only four of the fifteen cases had intelligence quotients over 85. (Table 156).

The schooling of the cases of ataxia is shown in Table 157. It will be seen that more than half of those over the age of five attended schools for the physically handicapped, a reflexion of the severe educational disability that ataxia imposes. This disability is partly the result of the slow manipulation which the patients show, partly the result of the difficulty they have in learning to recognise and form letters, and partly the result of the intellectual impairment.

Behaviour.

Four patients showed overactive behaviour of the type described in the section on hemiplegic paresis. In three the overactivity was marked and in one was less evident.

Three /

TABLE 156

The intelligence quotients of 15 patients with ataxia.

	115+	100-114	85-99	70-84	55-69	Under 55	Untested
Number of patients	0	1	3	6	3	0	2

TABLE 157

The schooling of 15 patients with ataxia

School	Number of patients
Normal schools.	2
Schools for the physically handicapped.	7
Schools for the mentally handicapped.	3
Ineducable.	0
Not yet at school.	3
Total	15

Case 72. Boy born in 1942.

Acquired ataxia and overactive behaviour of subacute onset and unknown aetiology. The first of two illegitimate boys born to a mother aged 17 at the time of his birth, and unknown father. Mother's brother is a psychopathic alcoholic.

Mother said to be well during pregnancy. Child born at term after 12 hour labour. Birth weight 7½ lbs. Normal early development. Walked and said first words before age of one year. From the age of two onwards he had chronic bilateral otitis media. From age of three had frequent seizures in which his face would become congested, eyes glazed, and he would foam at the mouth. He would become unconscious and stiff in trunk and all four limbs, with head retracted. He was incontinent in many of then. The attacka occurred as often as three or four times a day, apparently unrelated to any precipitating cause, for a period of six or seven weeks, and each would last from one to six or seven minutes. When they occurred as frequently as this, he was rather quiet, disinterested and inactive, sleeping for some hours after each attack. As they became less frequent, up to seven to ten times a week, he became more alert and active again.

Two years after the onset of attacks he developed whooping cough (1947) and seven weeks later was taken to hospital extremely ill, having as many as 42 paroxysms in a day, and in addition, showing superadded pneumonic complications. He required hospital treatment for four and a half months, during which no further convulsions occurred and none occurred following discharge.

Following this attack he seemed to alter in disposition, becoming increasingly active, restless and unable to keep still for a moment. His attention span diminished, and he ceased to be able to play with other children. He was liable to runabout himself, laughing and talking to himself for hours at a time. He was very temperamental and liable to burst into tears or have temper tantrums very readily. At the same time he seemed to become progressively unsteady and clumsy. He began to lean forwards as he walked and slouched, his shoulders pulled forwards. He began to fall frequently and to be uncertain on stairs. He could no longer run as quickly as his friends. Over a period of about seven months his unsteadiness increased, but then remained static and in the year prior to his being examined had improved somewhat. He was falling less, and was more confident on stairs. Activities which had become impossible for him on accouunt of his clumsiness, such as dressing himself and using a pencil, had been recovered. His overactive and disordered behaviour had not, however, improved.

On examination he was a small grossly overactive, unsteady /

unsteady jerky boy. He was never still, never quiet. He was continually moving about the examination room, picking things up and putting them down, talking all the time, and seeming quite unable to settle to anything. His attention span was very poor and sometimes he would wander away whilst listening to one or beginning to obey it. His speech was as rapid as his other activities, but there was no obvious dysarthria. There was no obvious asymmetry or wasting of the limbs. There was generalised hypotonia. This was more marked on the left than the right. There was bilateral hyperextensibility of the fingers. Power was within normal limits in all four limbs, and the range of voluntary movement was full. Co-ordination of all four limbs was impaired, intention tremor being very apparent on both finger nose, and hell knee tests, especially on the left. Fine movements were noticeably clumsy and fine repetitive movements were very badly performed.

The biceps, triceps, supinator knee and ankle jerks were all very sluggish, more so on the left than the right. The plantar responses were flexor, the abdominal reflexes were present.

When he walked, he did so on a broad base, swaying from side to side and banging his heel down as he proceeded, before the toes. In spite of this, he rarely moved more slowly than at a run, usually with his arms flexed at the elbows and flexed and elevated at the shoulders so that his hands were at the level of his nipples. When blindfolded he swayed more and deviated inconstantly to the left. The Romberg test was negative. No abnormalities of skin sensation, or joint or position sense could be detected.

TABLE 158

Cranial nerve involvement in 15 patients with ataxia

		Number of patients
Visual defects.		2
Strabismus	unilateral	1
	bilateral	4
Nystagmus.		2
Facial paresis	upper motor neurone	4
	lower motor neurone.	2
Deafness	unilateral	1
	bilateral	1
Tongue involvement.		1

Three of the patients showing overactive behaviour showed epilepsy. The overactivity was sufficiently severe in all the cases to make education by normal methods a very difficult and strenuous undertaking for the teacher.

Speech defects in ataxic patients.

It will be seen from Table 159 that 11 of the 15 ataxic patients showed speech defects. In 8 cases articulatory defects of varying degrees of severity were present, in four of the cases accompanied by scanning speech. Scanning speech without other abnormalities was evident in 3 cases.

Visual defects.

One patient with severe predominantly unilateral ataxia showed unilateral optic atrophy with complete unilateral amaurosis. The opposite eye was unaffected. One patient with bilateral ataxia of moderately severe degree and mild left hemiplegia showed complete left homonymous hemianopia, and some impairment of vision in the remaining half field in the lower quadrant bilaterally. One patient showed severe bilateral astigmatism.

Cranial nerve involvement in cases of ataxia.

These are shown in Table 158 .

Strabismus.

Unilateral abducent paresis was present in one case and bilateral abducent paresis in four patients. In the latter the abducent paresis was asymmetrical in severity in three of the patients and symmetrical in one. The patient with unilateral abducent paresis also had amaurosis of one eye.

No /

TABLE 159

Speech defects in 15 patients suffering from ataxia

Disorder of speech	No. of patients
Articulatory abnormalities.	4
Scanning speech.	3
Articulatory abnormalities and scanning speech	4
Total	11

No cases of third nerve involvement were encountered.

Nystagmus.

Two cases had nystagmus. One of these showed homonymous field defects, and nystagmus of a course type in all directions. She had bilateral moderately severe ataxia. The other case also showed moderately severe bilateral ataxia. Nystagmus was present only on gaze to the right and was slow and swinging in type.

Facial involvement.

Five of the 6 patients with mild hemiplegia associated with their ataxia showed facial paresis of upper motor neurone type. In none of the cases was there asymmetry at rest, but slight lag was evident on voluntary movement. In 4 of the cases this lag was also evident on emotional movement, but one showed overaction of the face on emotional movement, and lag on voluntary movement.

One patient showed facial paresis of lower motor neurone type which followed damage to the facial nerve suffered during meningitis.

Hearing.

One patient was totally deaf in one ear as a result of meningitis. One patient was totally deaf as a result of tuberculous meningitis treated by streptomycin. He also showed absence of vestibular response bilaterally on the caloric tests.

Swallowing.

None /

None of the ataxic patients showed apparent difficulty in swallowing at the time of examination.

Tongue.

One patient with moderately severe predominantly unilateral ataxia, slight hemiplegia and partial hemianaesthesia showed a tendency for the tongue to deviate to the right when central protrusion was attempted.

Epilepsy.

Three patients with ataxia showed grand mal epilepsy. In 2 the ataxia was predominantly unilateral, and in one, bilateral. One of the cases was severely affected, one moderately severely, and one slightly. One of the cases of grand mal also suffered from petit mal attacks.

One of the cases of grand mal had suffered from attacks once or twice a week from the time of birth to the age of 3, at which age they had ceased. The attacks had been accompanied by loss of consciousness, generalised twitching of the limbs and incontinence.

One girl with moderately severe predominantly unilateral ataxia, slight hemiplegia and hemianaesthesia following tuberculous meningitis showed grand mal attacks which occurred once every week on average. They were accompanied by loss of consciousness, incontinence and generalised twitchings of the limbs. The attacks lasted approximately 5 minutes on most occasions, but consciousness had been lost for as long as half an hour in some. The attacks were little affected by therapy. The patient with petit mal, in addition to grand mal /

mal showed attacks of the latter type over 3 - 4 months. They were severe in type with generalised twitching of the limbs and loss of consciousness for up to 3 hours. Her petit mal attacks occurred many times a day and were observed at one examination to have occurred 11 times in one and a half hours in spite of drug therapy.

One patient with moderately severe bilateral ataxia had seven attacks of epilepsy between the age of 14 months and 4 years which were of Jacksonian type and which started in his right hand, on the side affected by slight hemiplegia. Following the administration of phenobarbitone no attacks had been present for 18 months at the time of examination.

Hydrocephalus.

Two patients showed hydrocephalus of congenital type. In one, the hydrocephalus had become arrested spontaneously. The occipito frontal circumference was 25". One case had had third ventriculostomy and the head circumference was 2.13" at the age of 3.

Other findings.

One patient, without apparent visual defect showed a curious abnormality of the vessels in one retina. They were abnormally tortuous towards the temporal pole, almost angiomatous, but x-ray of the skull showed no abnormality and other angiomata were not discovered.

One patient had a congenital heart defect, thought to be a patent interventricular septum.

CHAPTER 10

The Literature on Dyskinesia in Childhood

Early Writers.

Occasional mention is made of involuntary movements in the affected limbs of children suffering from cerebral palsy in the earliest literature. Abnormal incoordinate finger movements occurring when hemiplegic patients attempted to use their affected hands were noted by Cazauvielh, (1827); Cotard, (1968); v. Heine, (1860); Charcot, (1853); A brief description of a microcephalic idiot in whom continual involuntary movements of the upper limbs were present was given by Delpech, (1828). "Les mains ne saisissaient les objets que d'une façon malhabile; elles ne pouvaient les maintenir un seul instant".

Little also described involuntary movements occurring in a number of his patients. At least one of them, (Case XLIII Arthur S.) was almost certainly a case of generalised choreoathetosis, (Little, 1862).

No one seems to have made systematic observations on the abnormal movements seen in children suffering from cerebral palsy until after 1871 when Hammond, in the first edition of his "Treatise on Diseases of the Nervous System", presented a description of "Athetosis", which he considered was a well defined disorder resulting from cerebral damage, the site of which was obscure though - "One probable seat of the morbid process is the corpus striatum".

The two cases he presented were both adults who had epilepsy /

epilepsy and showed persistent writhing movements of the fingers and toes on one side which varied in intensity from time to time but never ceased even during sleep. Voluntary movements of the affected limbs were much impeded by the presence of these movements because the antagonists tended to preponderate over the action of the agonists in both the hand and foot.

A reading of Hammond's original chapter leaves one in no doubt as to why the exact nature of the disordered movements he observed was so much debated by contemporary writers. The illustrations in his book suggest that they may have been merely extreme examples of the involuntary writhing movements found so commonly in hemiplegia. But his descriptions leave much to be desired as indicated by Gowers, (1876).

His work gave rise to great interest in the United States, Great Britain and in Europe. As a result of it many authors were stimulated to publish their observations on involuntary movements occurring in a variety of neurological disorders; it soon became apparent that the systematic description and classification of disorders of movement presented great difficulties.

In 1873 a further seven patients in whom involuntary movements were the major complaint were presented by Clay Shaw, under the title "On Athetosis or Imbecility with with ataxia". Unfortunately the histories, and in some instances, the clinical descriptions are very inadequate, but /

but it seems certain that the condition he was describing is that discussed by other authors under the heading of bilateral athetosis. Clay Shaw deliberately avoided this word because in his cases the involuntary movements had always been present, whereas Hammond's had been acquired sometime after birth as a result of disease. He notes "The disease most closely resembling it is chorea, but the distinction is, on a careful examination well marked ..... In the latter there is not the wavy gliding movement resembling the peristaltic activity of involuntary muscle and the jerkings are more sudden, rapid, and unexpected so that it is quite impossible to say which muscle will be next affected". He differentiated his patients "from others of the idiotic and imbecile class" and noted that the appearance of idiocy which was so common in patients suffering from " imbecility with ataxia" might be very misleading.

Raymond emphasised the very frequent association with hemiplegia of the movements described by Hammond and distinguished two types of involuntary movements, hemichorea and hemiathetosis. Hemiathetosis was a slow writhing movement found in the hands and feet, especially the fingers and toes, present most markedly on attempts at voluntary movement in patients who showed, or had previously shown some degree of hemiplegia. Hemichorea was distinguished from hemiathetosis by the fact that the movements were more rapid, less constant in type and affected the proximal rather than /

than the distal parts of the limbs. They were exacerbated by voluntary movement but were rarely completely abolished by rest though they tended to disappear during sleep. Raymond did not discuss bilateral athetosis or chorea as a form of cerebral palsy. (Raymond, 1876). A more detailed analysis of the form of disordered movement associated with hemiplegia, especially in childhood was presented by Gowers in the same year. (Table 60)

He emphasised that the description of "Athetosis" by Hammond was not specific enough for the condition to be considered as being more than a symptom of a number of diseases, the most common of which was hemiplegia. In this condition involuntary movements usually appeared as paresis receded. This was especially liable to occur in childhood. A variety of different forms of involuntary movements were described which could be graded, as in a spectrum, from the "quick clinic spasm of intermitting type" to "tonic spasm", which much more nearly resembled "Fixed rigidity".

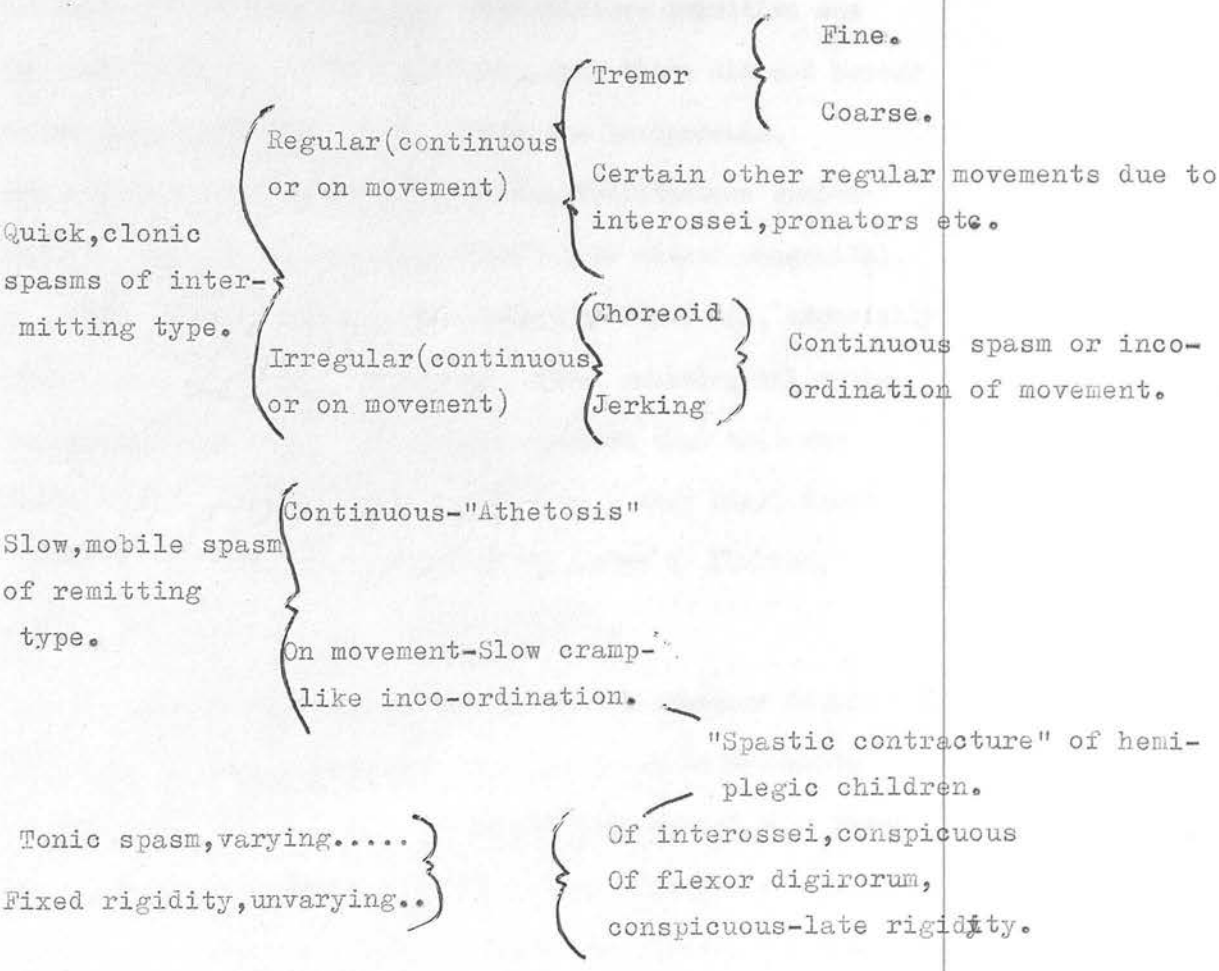
Further studies of post-hemiplegic movement disorders were presented by Eulenberg, (1889) and Bernhardt, (1895). Both these authors presented cases of acute hemiplegia occurring in early life in whom involuntary movements of the affected limbs became progressively apparent as the paresis waned.

Oulmont defined two forms of hémiathétosis. Hemiathetose symptomatique was the name given to the slow writhing /

Table 160.

Gowers, W.R. (1876)

Classification of post-hemiplegic disorders of movement.



writhing involuntary movements of the distal parts of the limbs when these were associated with hemiparesis or other evidence of cerebral damage. Hémiathétose primitive was the name given to similar movements when these did not appear to be associated with, or to follow the hemiparesis. Hemiathetose primitive was rarer than Hémiathétose symptomatique, but did occasionally result from either congenital, or, (more frequently) acquired damage to the brain, especially after acute infectious diseases. Other neurological manifestations were absent and Oulmont assumed that this was because the movements could result from a very small focal lesion in the brain as previously suggested by Charcot, (1853) (Thése de Paris). Oulmont, (1878).

In contrast to hémiathétose there was athétose double (athétose generale) which was the name given to movements of identical type to those of hémiathétose except that these were present bilaterally, and were often also present in the face. The condition usually occurred in idiots, but not invariably so and a few cases were reported in which the patients (usually children) had appeared to be of average intelligence. The involuntary movements had been present from shortly after birth in the vast majority of cases and a history of abnormal birth was often elicited. Oulmont presented a number of cases from the literature including the seven briefly recorded by Clay Shaw (1873). He added four more cases. A detailed reading of these suggests that Oulmont /

Oulmont adhered less strictly to his definition of athetoid movements occurring only in the distal parts of the limbs than he might. More difficulties in head control, involuntary jerking movements of the shoulders and upper arms are described in "Observation XXXI" in addition to writhing movements of the hands and feet. As in the cases of Clay Shaw it seems that the category of athétose double could include disorders in which movements reminiscent of chorea, rather than those of slow writhing quality which Hammond originally described as "Athetosis".

Following this work a large number of case reports of patients suffering from double athetosis appeared (Warner, 1881; Ross, 1882; Barrs, 1885; Adersen, 1886; Simpson, 1870). Osler discussed them as follows - "A class of cases belonging to this division of bilateral hemiplegia is characterised by spasm and disordered movement. They are described in the literature as chorea spastica and double athetosis. The cases I refer to are simple aspastic diplegias, plus post hemiplegic disorders of movement. The history is the same as in ordinary cases; the trouble has persisted from birth or shortly after and there is a condition of feeble-mindedness or idiocy, though in some instances the intelligence is fair. Very often there has been difficult labour". He then presented five very vivid and detailed case descriptions of typical cases. It is of interest that he was aware of the abnormal teeth so often found in patients with dyskinesia and also of the occasional occurrence of nystagmus. His case 18. is the first mention of jaundice /

jaundice in infancy being a finding of possible aetiological importance - "had jaundice when 11 days old after which the paralysis occurred". One particularly good report is the second case in "Athetosis and Athetoid Movements in the Insane", by Richard B. Mitchell (of Edinburgh) 1882.

On the basis of very detailed descriptions of involuntary movements found in a series of children who appeared to have suffered cerebral injury at birth, a category of chronic chorea of congenital origin was defined by Huet, (1889). Some of the cases he derived from the literature under this title are also referred to by Oulmont and Audry, (1892). Huet's thesis was of value in leading to a further understanding of the great differences in the type of involuntary movements found in neurological diseases in childhood.

Ninety-three cases of generalised athetosis (some of them doubtful) were collected from the literature and from his own practice by Audry, (1892). In "L'athétose double et les chorées chroniques de l'enfance", he reviewed the previous literature in great detail and concluded that involuntary movements in the limbs could be symptoms of a large number of diverse neurological conditions. On the other hand double athetosis in childhood was most often a congenital condition which follows abnormal labour or delivery and often failure to breath immediately after birth. Boys and girls were about equally often affected. The movements were more often evident within the first two or three /

three years of life. Their character and extent varied considerably from case to case. Most commonly they were found in the fingers, hands, toes and feet, but the face, eyes, tongue, muscles of the head and neck and of the proximal parts of the limbs might also be involved. The lower limbs were generally less affected than the upper. The character of the movements varied from being very slow and writhing to being almost choreic in type. Movements usually, but not always, ceased during sleep and were diminished when the patient was relaxed. They were much exacerbated by any voluntary movement of the limbs, especially the affected limbs. They tended to be more severe when the child was excited or emotionally tense, fatigued or cold. In some cases they were powerful enough to cause subluxations of the joints, especially the interphalangeal joints. Scoliosis was frequently found. The gait was commonly very unsteady and jerky, but not truly ataxic as Clay Shaw had suggested. Muscular atrophy was occasionally found in association with the involuntary movements, but in other cases apparent hypertrophy was present. Speech disturbances, commonly slow and inaccurate pronunciation, dissimilar to those found in ordinary (rheumatic) chorea, were the rule rather than the exception. Involuntary movements of the tongue were often largely responsible. Drooling of saliva was common and contributed to the idiotic appearance of many patients. Attempts at speech were usually accompanied by much grimacing. Intelligence was commonly, /

commonly, but not invariably impaired. Epilepsy was relatively uncommon compared to other forms of cerebral palsy. Sensory loss was rare. Associated paresis, spasticity and contractures, together with alterations in the reflexes, especially in the lower limbs were found in many patients. Mixed pictures of spastic diplegia and bilateral athetosis were sometimes seen. Autopsy findings were variable. Generalised cerebral atrophy and evidence of damage to the basal ganglia were the most frequent (as previously reported by Massalongo, 1890). Audry was unwilling to make any clear nosological distinction between congenital chronic chorea (Huet) and congenital bilateral athetosis (Oulmont).

Freud also considered congenital chorea and generalised athetosis to be closely related disorders from the symptomatological point of view. They differed merely in the type of involuntary movement which was predominant. Both were frequently found in association with spastic signs and intertypes linking "Tabes spasmodique" (spastic diplegia) on the one hand and generalised chorea or athetosis on the other, could readily be found. From the aetiological point of view, abnormal parturition was important. A history of apnoea after delivery was often found. Occasionally cases occurred following infectious diseases in early childhood, (Freud, 1893; 1897).

Thus, by the turn of the century, the existence of a class of congenital cerebral palsy in which involuntary movements /

movements were a more marked feature than actual paresis had been established. It was realised that a distinction had to be made between this group of cases in which the involuntary movements were almost always generalised and athetosis complicating hemiplegia. The clinical picture of generalised athetosis, or generalised congenital chorea, or choreoathetosis was variable, and that in some patients other forms of cerebral palsy might co-exist. Most of the autopsies on patients had revealed abnormalities in the basal ganglia, (Putnam 1892), though it was emphasised by some authors (especially Audry) that damage elsewhere in the brain or brain stem might produce a similar clinical picture. The aetiology was only partly known, but "the factors of Little" were acknowledged to be important, and Freud had laid special stress on the frequency with which apnoea after delivery was found in case histories.

More Recent Work: Aetiology and Pathology.

After 1900 the aetiological and pathological aspects of of the dynskinetic disorders attracted more attention than did the clinical findings which had already been so fully described. This trend was encouraged by the description in 1911 of "Progressive Lenticular Degeneration with Cirrhosis of the Liver" by Wilson and the current physiological work on postural mechanisms dependent upon the basal ganglia, (Walshe, 1947; Wilson, 1912).

Pathological knowledge of congenital choreoathetosis was /

was much advanced by the exhaustive and detailed work of the Vogts on "Status Marmoratus" and related findings in the basal ganglia. (Status marmoratus - the presence of myelinated fibres in aggregations of a density abnormal for that particular region of the nervous system, Norman, 1947). In the course of a long series of complex papers written in difficult German, these authors tried to define a "pure striatal syndrome" which was the result of damage confined to the basal ganglia most often congenital in origin. There was dysarthria accompanied by generalised "Hyperkinesis with involuntary movements of all four limbs, especially of choreic type and pathological (false) laughing and crying. Intellectual functions were usually well preserved. They considered that "Status Marmoratus", a lesion which they considered to be of prenatal origin, was the classical pathological finding, (Oppenheim and Vogt, 1911; Vogt, C. and Vogt, O. 1919, 1920, 1~~923~~). Rather similar attempts to correlate the distribution of pathological lesions in the basal ganglia with variations in the clinical picture were made by Jakob, (1925); Herz, (1944).

A survey of the clinical conditions encountered in children which could be attributed to diseases of the basal ganglia was made by Crothers (1921. He described one case of hepatolenticular degeneration (Wilson's Disease) and two atypical cases of deteriorating dyskinesia, as well as three patients with a history of birth injury in whom he suspected birth /

birth damage in the basal ganglia. He described the appearance and forms of the involuntary movements in considerable and lucid detail. He did not accept the Vogts' concept of congenital choreoathetosis being due to a congenital malformation of the basal ganglia, but emphasised the likelihood of birth injury.

The contemporary interest in the pathological aspects of birth injuries was reflected in increased research on the problem of the aetiology of congenital dyskinesia in childhood, (Ford, 1926; Holland, 1922). The distribution of the localised haemorrhages and cerebral softenings observed in fatal cases of birth injury had been shown to be often periventricular and especially liable to occur in the basal ganglia, (Schwartz, 1924, 1926; Marburg, 1936). The mechanism of the production of these lesions was debated. Some authors emphasised the action of generalised foetal anoxia during or immediately after birth as has been described by Courville, (1950, 1952). Others thought that obstruction due to kinking of the great cerebral vein in conditions of oblique stress on the cranium was a more frequent cause of venous stasis in the tributaries of the great cerebral vein and consequent localised damage in the basal ganglia, (Marburg, 1936; Ford, 1926; Malamud, 1950). ("Asphyxia obviously plays a supporting, but never a decisive role", Marburg and Casamajor, 1944).

Though there might be argument about the mechanism by which /

which damage resulted in the basal ganglia during the process of abnormal birth there was a strong body of opinion that birth injury was responsible for "Status Marmoratus", (Lowenberg and Malamud, 1933; Norman, 1944, 1947; Malamud, 1950; Benda, 1945).

The opinion of the Vogts that this condition was a congenital malformation has, however, also received support. It has been regarded as the underlying pathology of a variety of congenital syndromes of involuntary movements, some of which are hereditary and others progressive, by Alexander, (1942) and Denny-Brown, (1946). On the other hand, recent clinical surveys of children suffering from choreoid movements of congenital origin have tended to emphasise the importance of birth injury and particularly paranatal hypoxia in the majority of cases, (Byers, 1942; Asher, 1952; Evans, 1948; Phelps, 1951; Perlstein, 1952).

Kernicterus.

As early as 1875 it had been observed that babies dying of icterus gravis showed deep yellow pigmentation of the tissues of the nervous system, and particularly of the basal ganglia. Though the clinical details of his patient, a second born girl, are rather scanty, (and the family history non-existent), the neuropathological findings were clearly described in "Ueber das Vorkommen von Bilirubinkrystallen bei neugebornen Kindern", by Orth in 1875. Similar findings were reported by Schmorl (1903). This author examined the brains of 120 children dying as a result of icterus gravis and /

and found that six of them showed yellow staining in the nuclear masses of the brain. Other case reports with more detailed clinical histories followed quite rapidly, (Beneke, 1904, 1907; Pfannenstiel, 1908; Esch, 1908; Knopfelmacher, 1910; Yllpo, 1918; Thorling, 1922; Hoffman and Hausmann, 1926). As a result of these it became possible to define a clinical picture of "kernicterus" which was summarised as follows by Zimmerman and Yannet, (1933).

"1. The children are apparently normal at birth, labor usually being uncomplicated.

2. Jaundice appears early, usually before the second day and is intense.

3. Evidence of involvement of the central nervous system, such as convulsions and spasticity, is frequent.

4. Death usually occurs early, on or before the fifth day of life".

The neurological findings in affected children had been described in some detail by this time. The occurrence of generalised intermittent rigidity, most marked in the trunk and often associated with opisthotonos has been stressed by de Lange, (1924, 1925), Gruenwald and Messer, (1927). General poverty of movement, marked drowsiness or irritability in feeding, sometimes apparently the result of being unable to swallow, a high pitched cry, and occasionally nystagmoid eye movements had also been observed. Generalised convulsions were frequent. It was not unusual to have more than one /

one child in the family affected. There was great controversy about the ætiological factors involved in kernicterus. Some authors maintained that infection was the precipitating factor of kernicterus. They included Beneke, (1907), Esch, (1908), Schmorl, (1903), Riemond and van Creveld (1937). Others postulated functional immaturity of the liver and thought the condition analegous to Wilson's Disease, or the operation of toxins which were harmless to the mother but harmful to the child. That some haemolytic factor was transferred to the child by the mother, and that the cerebral damage might be due to accumulation of the products of haemolysis was suggested by Hoffman and Hausmann, (1926). But this suggestion did not answer the other questions raised first by Schmorl, (1903), as to whether the pigmented cells in the brain were pigmented because they had been damaged or killed, and therefore had an attraction for bilirubin, or whether they had been normal until damaged by the accumulation of the pigment itself.

The Rhesus Factor and the causes of Kernicterus.

The greatest step forwards in the understanding of the aetiology of kernicterus was the recognition of the rhesus factor by Levene et al, (1939). The history of the gradual increase in knowledge which finally resulted in this achievement has been fully reviewed by Potter, (1944). That a mother without this factor could make antibodies to it if the foetus was Rh positive, rapidly became apparent. (Wiener and Peters, 1940; Levine et al, 1940; Wiener, 1941). The importance /

importance of these observations in erythroblastosis foetalis was rapidly realised, (Levine et al, 1941a, 1941b). Indeed it appeared that the problem of the aetiology of kernicterus was almost solved for a number of years, (Claireaux, 1950). A number of authors stated, for example, that rhesus incompatability was the sole cause of kernicterus in the newborn, (Becker and Vogel, 1948; Wiener, 1946). This view was contested by Vaughan, (1946), Parsons, (1947) and by Zuelzer and Mudgett, (1950). The latter authors pointed out that "study of the aetiologic factors in 55 consecutive cases of kernicterus seen at autopsy disclosed the existence of a large seemingly heterogenous group in which erythroblastosis was not a causative factor". In 32 patients they found a higher incidence of first born infants, a higher percentage of negroes. Whilst some of these cases might be explained on the basis of ABO incompatibility, "prematurity, severe infections, diarrhoeal disease, pulmonary and cerebral haemorrhage and maternal diabetes were the chief conditions encountered in those cases in which erythroblastosis foetalis was excluded".

Kernicterus in 24 premature babies who showed no evidence of blood incompatibility with their mothers was reported by Aidin et al, (1950). The pathological picture was identical to that found in erythroblastosis foetalis and the clinical picture was dissimilar. Ten patients suffering from "Athetosis" who had suffered from kernicterus, but who were rhesus compatible with their mothers, were reported by Asher, (1952). /

Asher, (1952). A more detailed clinical account of kernicterus in the presence of rhesus compatability in premature babies was given by Govan and Scott, (1953). Since this time there has been much speculation about the various contributory aetiological factors of importance in kernicterus. Aidin et al, (1950) suggested that the effect of oxygen lack on diminishing the blood brain barrier might be important. The contributory effects of birth trauma and of a high level of circulating ammonia have also been much under discussion, (Leikin, 1957). There is some experimental evidence that bilirubin in high concentrations may itself be damaging to the nervous system, (Kuster and Krebs, 1950). But it is difficult to reconcile these observations with the fact that the majority of patients with atresia of the bile ducts do not show the clinical picture of kernicterus.

Of more immediate practical importance have been the advances made in the treatment of kernicterus since the end of the war. The technique of exchange transfusion through the umbilical vein developed by Allen and Diamond meant that in many cases of rhesus incompatability kernicterus could be prevented, (Diamond, 1947; Farquhar and Lewis, 1948). It is now extremely uncommon to find persistent neurological defects in erythroblastic children who have been adequately transfused by the exchange method in the first few hours after birth, (Allen and Diamond, 1957). The author has seen only two cases of choreoathetosis apparently attributable to kernicterus in whom adequate exchange transfusions seemed

to /

to have been performed in a series of almost 300 cases of dyskinesia in childhood.

The "post-kernicteric syndrome".

For many years, kernicterus was regarded as being an invariably lethal condition, though the possibility of infants surviving had been realised. The first case report is probably that of Osler, (1889) which has been mentioned, though he does not appear to have realised its possible significance. Single cases of survival after what appeared to have been kernicterus were published by Morris, (1900), Arkwright, (1902), Pitfield, (1912), Guthrie, (1913). Four patients, one of whom was delivered prematurely, were described by Spiller, (1915). He notes that in each case "the jaundice was believed by the parents to be responsible for the child's condition". Spiller diagnosed his cases as suffering from spastic diplegia, but in fact they all showed marked choreoid and athetoid movements, and there is little doubt that they would now be called cases of congenital choreoathetosis. Hoffman and Hausmann, (1926), added two more well described cases in whom clinical indications of kernicterus were followed by mental retardation and motor incoordination. Eleven cases of recovery with residual damage were collected from the literature and a further typical case was contributed by Greenwald and Messer, (1927).

On the basis of these reports and their own experience of children in a colony for defectives, Zimmerman and Yannet could /

could present a much more detailed clinical picture of the effects of kernicterus in later life. (Zimmerman and Yannet, 1933). There was a triad of symptoms, namely, "a severe neonatal illness associated with jaundice, subsequent evidence of damage to the basal ganglions, and a familial history of icterus gravis". The evidence of damage to the basal ganglions (sic) consisted of delayed motor milestones, motor incoordination, "extrapyramidal spasticity, athetoid and choreiform movements and emotional instability". Detailed case histories of two more survivors were presented.

In part, as a result of increased interest in the condition, and in part due to better treatment, increasing numbers of survivors of kernicterus were reported. Five more (one of whom was noted to be deaf) were described by Coquet, (1944). She attempted to differentiate the neurological picture found after kernicterus from that found in "generalised athetosis". She felt that in the latter condition contractures of the limbs, hemiparesis, and persistent alterations of muscular tone occur much more commonly. Whether tonic states ("extrapyramidal spasticity") or choreoid movements predominanted in cases following kernicterus depended upon whether pallidonigral or putaminocaudate lesions were the more marked. She thought the underlying lesion was the "état dysmyelinique of the Vogts". The relationship of the pathological findings to the clinical picture of patients showing extrapyramial symptomatology was further discussed by Docter and Fitzgerald, (1945) and Greenfield and Kounine, (1939).

Leude /

Lande added deafness, transient blindness which usually recovered within the first two years of life, and irregular eye movements to the clinical picture of generalised athetosis or choreoid movements, persistent or intermittent extrapyramidal or pyramidal increase of tone, ataxia, disorders of balance, or atonic diplegia considered to be characteristic of the after-effects of kernicterus, (Lande, 1948). Perlstein stressed that the appearance of involuntary movements might be delayed for as long as a year after birth in some patients. He found some degree of deafness, usually of the high tone type in 40% of patients suffering from cerebral palsy after kernicterus. "Auditory aphasia" was often present. Most patients were unable to elevate or depress their eyes fully. In three quarters of the patients the intelligence was unimpaired, (Perlstein, 1950). Paralytic deafness, usually of high tone type was described in 16 of 22 children who had suffered from kernicterus, (Crabtree and Gerrard, 1950). Some indication of the importance of kernicterus in causing involuntary movements in childhood was given by the finding that 34 of 63 "athetoid" (dyskinetic) patients had a history of jaundice in infancy, (Asher, 1952). This author found that 22 of 24 patients who had been jaundiced in the neonatal period had some degree of loss of hearing and only four of 18 non-jaundiced cases.

The follow-up of 23 cases of kernicterus by Byers Paine and Crothers is probably the most comprehensive (and one of the briefest) descriptions of the clinical syndrome of

"Extrapyramidal /

"Extrapyramidal cerebral palsy with hearing loss following erythroblastosis", (Byers et al, 1955). They emphasise the changing clinical picture at different ages. Between the second and the fifth days there are the signs of kernicterus, severe jaundice with generalised hypotonia, bloody nasal discharge, and opisthotonos, poor Moro reflex, high pitched cry, poor sucking and frequent vomiting. Convulsions occasionally occur in the newborn period, but uncommon later. From the age of 3 to 6 months to perhaps 2 years, the characteristic picture is one of marked retardation of motor development and hypotonus accompanied by lively tendon reflexes and persistent immature postural patterns such as the tonic neck and neck-righting reflexes. Sometimes as early as 18 months, but more often later, the onset of involuntary movements occurs. The degree to which patients are disabled varies greatly.

Hand use is often severely impaired. Dysarthria "of the characteristic extrapyramidal type" is frequent. Grimacing and drooling are often accompanied by difficulty in chewing and swallowing. Tension, an intermittent increase of tone in the limbs, exacerbated by any attempt to move, is common. The Babinski and Hoffman signs may be positive. Moderate or severe hearing loss was definite in twelve of the nineteen patients who were tested. Only four of the fifteen fully tested patients were found to have intelligence quotients below 70. Restriction of eye movements, especially difficulty in convergence, and therefore also in looking upwards /

upwards was often found.

The teeth in children who had suffered from kernicterus had often been noted to be bad and green staining, especially of the incisors had been observed by a number of authors. A more detailed examination of the reasons for the premature decay and liability to caries of the teeth after kernicterus was carried out by Forrester and Miller (1955). These authors noted that there were enamel defects in these patients and pointed out that the level on the tooth at which this occurred was some indication of the timing when the damage to the enamel organs was sustained.

The Therapists.

Whilst the interest in cerebral palsy until the 1930s had been predominantly academic, and very little had been written on treatment, thereafter the growth of therapeutic interest and of educational and medical services for the child with cerebral palsy had its effect. Many of the early schemes of treatment were directed to the more intelligent physically handicapped child. Since a relatively greater proportion of children with dyskinesia retained intelligence than did patients with other forms of cerebral palsy they were closely studied, (Klingman and Carlson, 1937; Phelps, 1941, 1943; Byers, 1942; Evans, 1948). This resulted in increasingly detailed study of their symptoms, neurological signs and possible techniques of treatment which might be used to help them, (Phelps, 1942; Carlson, 1942; Crothers, 1942). Increasingly fine distinctions between the various

forms /

forms of involuntary movements were made and attempts, mostly haphazard, to relate the variations observed in the clinical pictures amongst the group of dyskinesias to pathological differences. The most detailed system of classification proposed appears to have been that of Phelps, summarised by Hellebrandt, (1950-51).

The characteristics of the condition are listed as follows -

"1. Slow purposeless involuntary movements.
2. Agonists and antagonists involved without regard for their reciprocal use.
3. The athetotic pattern is superimposed upon essentially normal voluntary effort.
4. Muscles have normal tone at rest.
5. No stiffness except when resistance is offered to the athetoid train.
6. Athetosis may be overcome in part by tension. (Thus the patient may simulate spasticity or rigidity). Tension is non-intentional and generalised.
7. Deep reflexes are essentially normal. May be reinforced by tension.
8. Superficial reflexes normal.
9. Secondary incoordination due to athetosis.
10. Overflow may be widespread.
11. Irritability and contractibility are essentially normal.
12. Deformities with contractures are relatively uncommon."

It will be observed that these features of the condition are not those which would immediately allow of a satisfactory neurological definition of "Athetosis". On the other hand, by concentrating on the alterations of muscle tone and movement patterns which are obvious to a therapist, a recognisable picture of disordered function is presented. The various variations encountered in the clinical picture are further represented in the following:-

Subtype of Athetosis

"

1. Emotional Release Athetosis. Labile response to sensory stimuli and affective states.
2. Cerebellar Release Athetosis. Exaggerated balancing and adjustment responses as though walking on a tight rope.
3. Shudder Athetosis. Involuntary mycoclonic-like response which may be sudden and violent.
4. Tremor Athetoid; single plane involuntary movement with rhythmical alternate contraction of agonist and antagonist.
5. Rotary Athetoid. Involuntary movements occur primarily around longitudinal axis so that flexion extension patterns are defective.
6. Hemi-athetoid. One sided athetosis.
7. Arm and Neck Athetosis. Excessive tension and athetosis in the head, neck and arm regions with relatively good lower extremities and at times "educated feet".
8. Deaf Athetoid; hearing loss with high pitch cut-off and loss of overtones; secondary speech defect due to hearing loss; spontaneous lip readers; weakness of upward eye movements; usually quadriplegic with relatively little involvement of the legs. "I have yet to see one who did not learn to walk". Phelps. Usually bright-eyed and alert with normal I.Q.
9. Dystonic Athetosis. Relatively little motion but characterised by by the assumption of bizarre and distorted positions which may be held for a considerable time.
10. Tension Athetosis; high degree of muscular tension is the major characteristic;trigger point release is common.
11. Non-tension Athetosis; limbs are flabby; do not resist passively produced motion; limbs may show a startling degree of flaccidity between athetoid trains.
12. Flail Athetosis; involuntary motion limited primarily to the hip and shoulder regions, extremity as a whole is thrown like a flail; peripheral parts are relatively uninvolved; arm may move like the vanes of a windmill. "

The complexity, nosological confusion and overlapping of categories within this system makes nonsense of it as a classification, but as a description of the various patterns of involuntary movements which are encountered it appears to be compatible with clinical experience.

The extent to which immature reflex patterns characteristic of the first weeks of life may persist in children with dyskinesia has been described by a number of authors, (Bobath and Bobath, 1954; Byers, Paine and Crothers, 1955).

In severe cases the Moro response may be elicited even at the age of four or five years and persistence of the symmetrical neck and tonic neck reflexes is highly characteristic of the congenital extrapyramidal disorders. Attempts to use these reflexes in initiating smooth voluntary movement patterns which will tend to diminish the effects of involuntary movements have been widely publicised, and are of great theoretical interest. Thus Temple Fay described homolateral and reciprocal crawling patterns which are based on the swimming movements found in the first weeks of life ("Amphibian and Reptilian movements") and are used in an attempt to obtain smooth limb flexion and extension. More stress is put on the importance of full trunk and neck flexion to inhibit excessive tension in muscles by the Bobaths. Most physiotherapists wittingly or unwittingly use or adapt the tonic neck reflexes to their purposes in these cases, (Thomson, 1955).

Unfortunately it has not been possible to correlate the clinical pictures described by Phelps to pathological or aetiological entities except in the case of "The Deaf Athetoid" whose disorder appears to follow kernicterus in the vast majority of instances.

Recent Surveys./

Recent Surveys.

A number of recent surveys have helped to give some indication of the prevalence of dyskinesia and of visual, auditory and mental abnormalities in the condition. Different criteria of diagnosis are used by different authors for what is usually termed "Athetosis". For example, patients showing involuntary movements in the limbs as their main clinical abnormality are called "Athetoid" by Skatvedt, unless they show complicating rigidity (which is common), when they are placed in the category of "Mixed Cerebral Palsy". Lack of awareness of the dystonic phenomena of early diplegia has certainly led other authors to include patients in this category under the heading of "Athetosis" or "Dyskinesia", (Woods, 1957; Illingworth, 1958). In addition to differences in classification there are also differences in the population studies by different workers. As has been noted, series of patients referred to special schools for patients suffering from cerebral palsy are likely to contain a relatively high proportion of dyskinetic children, as they tend to be of higher intelligence than those suffering from other forms of cerebral palsy. The proportion of dyskinetic patients in regional surveys is distinctly lower. Thus, Phelps considers that 40%, and Illingworth that 25% of children suffering from cerebral palsy are "athetoids", (Phelps, 1951; Illingworth, 1958). Regional surveys suggest that between 10 and 20% of patients suffering from cerebral palsy have involuntary movements as their presenting clinical feature. /

feature. Thus 12% of 295 congenital cases were classified as "athetoid" by Asher and Schonell, (1950). Andersen found 17%, Woods 10%, Skatvedt 15% of unselected cerebral palsy patients to be "athetoid", (Andersen, 1954; Woods, 1957; Skatvedt, 1958). The prevalence of dyskinesia in the community is probably in the region of one in every two thousand births.

The majority of recent authors agree that there is a very high incidence of abnormal parturition with neonatal signs of birth injury, or of kernicterus in the histories of dyskinetic patients. In Wood's series only three of 33 children were the result of apparently uncomplicated pregnancies and deliveries, and 16 were severely jaundiced in the neonatal period, (Woods, 1957). Birth injury was considered to be more important in "athetosis" than in "Spastic Paralysis" by Asher and Schonell, (1950) and Skatvedt, (1958).

How deceptive the apparent mentally defective appearance of the vacant looking, drooling, grimacing, helpless, dyskinetic child may be has been emphasised by Phelps, (1941). The majority of authors agree that the child with dyskinesia is more likely to be intelligent than the patient suffering from other forms of cerebral palsy. Though relatively more physically handicapped, they more often retain their intelligence. Of 33 cases studied by Woods, 4 attended ordinary schools, 17 a school for intelligent children suffering from cerebral palsy, and two schools for the physically handicapped or deaf, (Woods, 1957). 23 of 31 dyskinetic /

dyskinetic patients were found to be of average intelligence by Skatvedt (1958). On the other hand, Asher and Schonell found that the mean intelligence quotient of "athetoid" and children with "bilateral spasticity" was comparable, (Asher and Schonell, 1950).

Epilepsy is found to be less frequent than in other forms of cerebral palsy in most series. The prevalence of seizures amongst dyskinetic children is variously quoted as being 4%, 7.5%, 15%, 27% and 32%, by Asher and Schonell, (1950), Skatvedt, (1958), Perlstein and Hood, (1956), Hopkins Bice and Colton, (1955), Illingworth, (1958). In all these series the prevalence of epilepsy amongst hemiplegic and "spastic quadriplegic" children is higher than the figures quoted for dyskinesia.

The electroencephalogram was noted to be abnormal in 20 of 35 "athetoid" patients studied by Skatvedt. In 8 there were focal features, in eleven generalised dysrrhythmia and bilateral abnormalities were noted in two, one of whom showed other abnormalities, (Skatvedt, 1958).

Impairment of hearing, amounting in some patients to virtual deafness, is found more frequently in dyskinesia than in other forms of cerebral palsy. The majority of hearing defects occur in patients who have suffered from kernicterus. 30% of dyskinetic patients were found to suffer from hearing impairment by Perlstein, (1953). Hearing was noted to be significantly defective in almost 20% by Asher and Schonell, (1950). /

(1950). Twelve of 33 patients were noted to have some loss of hearing by Woods, (1957), and 14 of 53 by Skatvedt, (1958).

Impaired visual acuity is apparently uncommon, but difficulties in visual fixation and in ocular movements are present in a large proportion of patients suffering from dyskinesia. Strabismus was observed in 17 of 33 patients by Woods, (1957).

Speech defects are common, not only because of the frequent impairment of hearing, but also because there is so often incoordination of articulatory and respiratory movements. 28 of 33 patients were noted to have defective speech by Woods, (1957). 27% had no speech, and 25% dysarthric speech in the series of Asher and Schonell, (1950). 66% of dyskinetic patients were noted to be dysarthric by Skatvedt, (1958).

Family History of Patients with Dyskinesia (Table 169)

The large majority of the parents of patients with dyskinesia were healthy. In Case 54. the father suffered from attacks of "Asthmatic bronchitis" which meant that he lost a certain amount of work each winter. The mother in this case suffered from anxiety neurosis and was an extremely excitable person, whom the observer suspected of having thyrotoxicosis on first meeting her. A brother of the patient in this case was said to have one abnormal foot on which he had been unable to wear an ordinary shoe during childhood, or an army boot later. It proved impossible to discover any more details about him. The only other abnormal sibling was in Case 125. in which a younger brother suffered from tuberculosis.

In Case 112. the mother suffered from a persistent facial weakness following an attack of Bell's palsy some years before. In Case 194. the mother suffered from chronic nephritis which was exacerbated by both her pregnancies. In Case 202. the father had healed tuberculosis of the cervical glands.

Only one relative was reported as being abnormal. The maternal uncle in Case 202. had died in early childhood from hydrocephalus.

Thus, a positive family history of neurological, mental or psychiatric disorder was relatively uncommonly obtained amongst patients suffering from dyskinesia. (Table 153).

TABLE 169

Family history of patients suffering from dyskinesia

Case No.	Social Class	Age of mother	Health of father	Health of mother	Unhealthy siblings	Relatives	No. of Preg.
21	IV.	32	Good.	Good.	None.	None.	1 of 1
23	IV.	24	Good.	Good.	None.	None.	1 of 1
37	III.	24	Good.	Good.	None.	None.	2 of 2
54	V.	30	Asthma. Bronchitis.	Anxiety. Nervous.	1 had abnormal foot.	None.	5 of 5
68	I.	23	Good.	Healthy.	None.	None.	1 of 2
106	II.	27	Good.	Healthy.	None.	None.	2 of 2
112	I.	35	Good.	Bell's palsy.	None.	None.	1 of 1
125	III.	36	Good.	Healthy.	1 Tuberculous.	None.	6 of 6
127	III.	45	Good.	Healthy.	None.	None.	3 of 3
136	II.	34	Good.	Healthy.	None.	None.	3 of 3
145	I.	44	Good.	Healthy.	None.	None.	3 of 3
146	II.	26	Good.	Healthy.	None.	None.	2 of 2
173	III.	26	Good.	Healthy.	None.	None.	1 of 3
178	II.	38	Good.	Healthy.	None.	None.	1 of 1
194	III.	25	Good.	Chronic nephritis.	None.	None.	2 of 2
197	V.	25	Good.	Obese.	None.	None.	3 of 3
202	III.	32	Healed tuberculosis.	Healthy.	None.	Maternal uncle hydrocephalic. 2 cousins tuberculous.	6 of 6

History of the other pregnancies to mothers of patients with dyskinesia.

There was a total of 46 pregnancies, including those with the patients, to the seventeen mothers of children suffering from dyskinesia. This gives an average of 2.7 per mother, which compares to that of diplegia (2.8 per mother) but is distinctly lower than that for acquired hemiplegia (3.8 per mother). (Table 161)

Of the 29 pregnancies, other than those resulting in the birth of the patients, to the 13 mothers with more than one pregnancy, two ended in abortion, both in Case 202. There were no stillbirths, but prematurely delivered twins died shortly after birth in Case 54. (the only case in which there was a possibly abnormal sibling), and there were two post-neonatal deaths in Cases 145 and 197. respectively. One child, again in Case 54. died at the age of 13 months from bronchopneumonia, though he had previously seemed quite normal.

Thus, in a total of 29 other pregnancies, twenty-four resulted in the birth of children who survived the first year, and twenty-two continued to survive and seemed normal at the time of examination, approximately 76%. This is a relatively high proportion compared to other forms of cerebral palsy, e.g. 54% of healthy survivors from other pregnancies of mothers with patients suffering from ataxic diplegia. (Table 162)

Obviously the causes for this may lie in better environmental conditions lessening the incidence of foetal and infant mortality, or they may lie in the fact that fewer abnormal children, /

Case No.	Social Class	Age of mother.	No. of preg. with patient.	Misc.	Other abnormal parturition.	Other normal.	Still-births.	Neonatal deaths.	Post neonatal deaths.	Abnormal siblings	Normal siblings.
37	III.	24	2nd. of 2	0	-	1	-	-	-	-	1
54	V.	30	5th. of 5	0	Prem. labour.	3	-	Prem. twins.	-	1 abnormal foot.	1, and dead(13/
68	I.	23	1st. of 2	0	0	1	-	-	-	-	1
106	I.	27	2nd. of 2	0	0	1	-	-	-	-	1
125	III.	36	6th. of 6	0	0	5	-	-	-	-	5
127	III.	45	3rd. of 3	0	0	2	-	-	-	-	2
136	II.	34	3rd. of 3	0	0	2	-	-	-	-	2
145	I.	44	3rd. of 3	0	0	2	-	-	1 "Respiratory infection"	-	1
146	II.	26	2nd. of 2	0	0	1	-	-	-	-	1
173	III.	26	1st. of 3	0	0	2	-	-	-	-	2
194	III.	25	2nd. of 2	0	1 chronic nephritis. and pre-eclampsia.	0	-	-	-	-	1
197	V.	25	3rd. of 3	0	1 Precipitate labour.	1	-	-	1 "pneumonia"	-	1
202	III.	32	6th. of 6	2	1 Breech delivery.	2	-	-	-	-	3

Table 162.

Fate of the other pregnancies of mothers with dyskinetic patients.

		Approx.%.
Total number of pregnancies.	46	-
Total number resulting in birth of patients.	I7	-
Total other pregnancies.	29	I00
Number ending in abortion.	2	7
Number otherwise abnormal.	4	I4
Number ending in stillbirth.	0	0
Number ending in neonatal death.	I	4
Number ending in postneonatal death.	2	7
Number of survivors at end of one year	24	83
Number of healthy survivors at time of examination.	22	76.

children, or children who are vulnerable to die from environmental effects are produced. A combination of these factors could be present.

Of the 29 other pregnancies only six were complicated, including the 2 ending in miscarriage which have been mentioned. In Case 54. there was an unexplained onset of premature labour in a twin pregnancy which resulted in the deaths of both infants almost immediately after delivery. In Case 194. there was preeclampsia in addition to underlying chronic nephritis. Labour was precipitate in Case 197. both with the patient and one older sibling "I just managed to get up the stair and had the fat wee bugger on the carpet". In Case 202. delivery of a previous child had been by the breech at term without other untoward occurrence. Taking account of abortions, but irrespective of other outcome, approximately 80% of other pregnancies were uncomplicated. This compares favourably with the 59% of other pregnancies which were normal in the group of ataxic diplegias. (Table 162)

Spacing of pregnancies.

Owing to the fact that as many as 14 of the 17 patients were the result of the last recorded pregnancies of the mothers the amount of information which could be derived from the spacing of pregnancies was limited, only two averages being available; that for the lapse of time between the birth of previous siblings, and that for the lapse of time between the immediately previous sibling and the patient.

Average lapse of time between delivery of 20 previous pregnancies. 2.9 years.

Average lapse of time between delivery of previous child and patient (11 cases) 3.7 years.

Thus, there is the same difference in the lapse of time between the births of previous children (or miscarriages) and the birth of the immediately previous child and the patient, as has been observed in other forms of cerebral palsy.

Number of pregnancy.

The place in the family and number of pregnancy is shown in the Table. 163

Table 163	In family	Pregnancy	
1st. and only	4	4	6
1st. of a number	2	2	
2nd. of a number	6	4	
3rd. of a number	3	4	
4th. of a number	0	0	
5th. of a number	0	1	
6th. of a number	2	2	

It /

It is of interest to note that as many as 14 of the 17 patients with dyskinesia were last born, at least at the time of examination, a very high proportion compared to the other forms of cerebral palsy described. This is probably related to the higher age of mothers in this group of cases, but possibly also to the deliberate restriction of pregnancy following grossly abnormal labours with the patients which are prevalent in this group. It is also interesting that almost a quarter were only children, again probably a consequence of the relatively high maternal age.

CHAPTER 11 b.

The Birth Histories of Children with Dyskinesia.

The birth histories of children who suffered from dyskinesia are summarised in Table 164 In 16 cases the disorder appeared to be of congenital origin, in the sense that the patients never gave convincing evidence of being normal for any prolonged period after birth. The exception, the girl in Case 125. appeared to be normal until the age of four years, and almost certainly acquired her disorder well after the neonatal period. The birth history was uncomplicated.

In 5 cases there were no overt clinical abnormalities of parturition, Cases 21, 54, 106, 112, and 145, though in Case 112. a cervical polypus was removed at the third month of pregnancy, and in Case 173. the mother was vaccinated at about the third month of gestation. It is of interest that three of these patients became jaundiced after delivery, and all of them were later shown to have been rhesus incompatible with /

TABLE 164

Birth history of patients with dyskinesia and their neonatal state

1. Congenital cases

Case No.	Abnormality of parturition	Immediate neonatal state	Bth.Wgt. lbs.	oz.	Subsequent neonatal course	Subsequent neurological abnormalities	Probable aetiology
21	Premature rupture of membranes.	Prolonged apnoea. Asphyxia pallida.	6	8	"Cerebral", irritable, restless and hypertoxic 3 days after birth.	Focal convulsions from 6 wks. of age.	Congenital.
23	Slight antepartum haem. Precipitate 2nd. stage. Cord round neck. (Mother Rh + ve)	Apnoea. Cyanosed. Slow to cry. Anaemic looking. Hyperpadis. Premature.	4	-	Rectal haem. 10 days. Petechiae within 12 hrs. Jaundice within 36 hrs. requiring 2 transfusions in 1st. week.	Sleepy lazy baby. Retarded milestones.	Congenital ABO incompatibility not excluded.
37	Prolapse of cord.	Prolonged apnoea. Regular respiration after 140 mins.	7	2	Restless, very still and feeble. Hypertoxic.	Went stiff in arm from 4-5 mths. Retarded milestones.	Congenital. Probably birth injury.
54	Normal. (Mother Rh - ve)	Normal. Cried at once.	6	4	Became jaundiced within 6 hrs. jaundice persisted for 3 wks. Then very pale.	Very placid baby. Jerky movements from age 1 month.	Congenital. Rh. incompatability.
68	Antepartum haem. after fall on day of onset of labour. Prolonged 1st. stage.	Normal.	6	13	Very placid lazy baby.	Clumsiness from age 6 mths. Unsteady from 1 year.	Congenital.
106	Normal. (Mother Rh - ve)	Normal.	8	8	Jaundiced at age 3 days for 6 wks. Drowsy. Hypotonic.	Retarded milestones.	Congenital.
112	Premature onset of labour 32 weeks. (Mother Rh - ve)	Prolonged apnoea. Cyanosis.	5	9	Jaundiced from 3rd. day for one month.	Jerking movements from 6 mths. Retarded milestones.	Congenital. Probable
127	Difficult forceps delivery.	Prolonged apnoea. Artificial respiration.	12	0	Very sleepy but irritable. Hypertoxic.	Jerky movements from age 10 months.	Congenital. Probable birth injury.
136	1 week post mature. Diff. high forceps at second attempt.	Fronted right clavicle. Cephalhaematoma. Much bruising. Cried at once.	7	8	Generalised convulsions at 12 hours.	Jerky movements of trunk and limbs for 3 mths. Retarded milestones.	Congenital. Probable birth injury.
145	Labour of 2 hours.	Normal.	7	2	Irritable, very floppy baby.	Jerky movements in trunk from age 5/12.	Congenital.
146	Postmature, 43 weeks. (Mother Rh - ve)	Normal.	7	8	Jaundiced within 24 hours and remained so for weeks. Required transfusions.	Floppy baby with abnormal movements at 10 mths. (in hospital until then).	Congenital. Probable kernicterus.
173	Foetal distress during labour.	Prolonged apnoea. Cyanosed. Slow response to resuscitation. "Club feet and hands".	6	8½	Sleepy baby. Poor feeding.	Jerky, clumsy movements. Retarded milestones.	Congenital. Probable birth injury.
178	Labour 2 weeks early. Prolonged high forceps.	Cried at once but then required resuscitation. Bruising over head.	8	12	Much respiratory difficulty. Hypertoxic. Could not suck.	Restless and "jumpy". Backwardness of development.	Congenital. Probable birth injury.
194	Chronic nephritis. Pre-eclampsia. (Mother Rh - ve)	Normal. Breathed at once.	8	3½	Much jaundiced within 24 hrs. Persistent until 6 weeks. Floppy.	Jerking attacks of trunk at 4 months of age.	Congenital. Kernicterus.
197	Onset of precipitate labour at 38 weeks. (Mother Rh - ve).	Jaundiced but cried at once.	5	4	Progressive jaundice 5 days. Persistent for 6 weeks.	Drowsy until age of 3 months. Jerky movements noted at 6 months.	Congenital. Kernicterus.
202	Premature labour. Prolonged 2nd. stage, 29 hrs. (6th. pregnancy). Breech extraction. Forceps.	Brief apnoea. Cleft palate and lip.	4	4	Good progress. But sleepy hypertoxic baby.	Attacks of stiffness at 9 weeks of age.	Congenital.

2. Acquired case

125	Normal.	Cried at once.	7	3	Normal progress.	Well until age 4 when fall from 1st. storey window. Twitching attacks thereafter. Progressive unco-ordination.	

with their mothers.

In 2 cases, 146 and 194, pregnancy only was disordered. In Case 194 the mother suffered from chronic nephritis and moderately severe pre-eclampsia necessitating admission to hospital a week before the onset of labour. Delivery was uncomplicated, and the child breathed at once, but later became jaundiced ..... due to erythroblastosis foetalis is was subsequently shown. In Case 146 there was post maturity and the mother and child were later found to be rhesus incompatible.

In seven cases, labour or delivery only appeared to be abnormal. In Case 37, labour was rather prolonged, lasting over thirty hours and the cord prolapsed. The child was born in a state of white asphysia, and had prolonged apnoe. In Case 197. labour was precipitate and occurred at an estimated 38 weeks gestation. In Cases 136, 178 and 202, labour was prolonged, being terminated by a successful high forceps delivery after a failed forceps delivery in Case 136; by a difficult high forceps delivery in the occipito posterior position in Case 178. and by breech extraction with forceps extraction to the after-coming head in Case 202. In Case 136. the child showed evidence of considerable trauma, but was not apnoeic, nor was the child in Case 178. though in Case 202. the premature baby showed apnoea and a cleft lip and palate. In Case 127. the child was extracted by a difficult forceps delivery by the General Practitioner after a labour of only four hours during which pains had been severe. The difficulties of labour and delivery were explained when it was found

that /

that the child's birth weight was twelve pounds. He was born in a state of blue asphyxia, and thought to be dead, but responded to injections of intracardiac coramine and lobeline and breathed in 20 minutes spontaneously.

In Case 173. foetal distress was noted during labour, the pulse slowing to under 80 per minute. The mother was immediately placed in the squatting position and the child delivered in a matter of seven minutes spontaneously. The baby was severely apnoeic at birth, however, and was noted to show "club feet and hands".

In two cases both pregnancy and labour, or delivery were abnormal. In Case 23. the single 18 year-old Irish mother had two small vaginal haemorrhages at an estimated 38 weeks' gestation. Her father insisted that she take a tram to the Maternity Hospital in spite of the fact that labour ensued quite rapidly and the child was delivered precipitously after a second stage of only 15 minutes after a first stage of 18 hours. The cord was round the child's neck, and he was severely apnoeic at birth and showed hypospadias. He became jaundiced within 36 hours.

In Case 68. ante partum haemorrhage occurred at an estimated 38 weeks' gestation when the mother fell from a ladder which then fell on top of her. She went into labour some hours later and after a prolonged labour of thirty-six hours was delivered spontaneously of a male child who breathed at /

at once and seemed normal.

The immediate neonatal condition and neonatal course of patients with dyskinesia.

In Table 164 are shown the clinical details of the immediate neonatal state and the neonatal course of the seventeen patients with dyskinesia. Seven were considered to be normal, including the patient with acquired dyskinesia who will henceforward be excluded from consideration. Three of these patients, Cases 54, 106, 145 were born after apparently uncomplicated pregnancy and delivery. In Case 194 there was preeclampsia superimposed on chronic nephritis, though labour and delivery were uncomplicated. In Case 68 there was antepartum haemorrhage and rather prolonged labour. In Case 46 there was post maturity.

There was apnoea in 8 of the remaining 10 patients. This occurred immediately after birth in 7 patients and after a few breaths in 1 case (178) who had been delivered by high forceps. The apnoea was considered to be severe, being prolonged for more than ten minutes in 6 cases (21, 23, 37, 142, 127 and 173) and mild in 2 cases (178 and 202). In 4 of the cases in which prolonged apnoea was present there were abnormalities of pregnancy, labour or delivery. But in two, parturition had apparently been normal apart from premature onset of labour in Case 112. In both cases with apnoea of shorter duration there were abnormalities of parturition.

The other abnormal infants were in Case 197, in which jaundice /

jaundice was noted immediately after birth, and Case 136 in which a fractured clavicle, a large cephalhaematoma and much head bruising was evident, though both the infants cried immediately after birth.

Irritability, generalised hypertonicity of the limbs and convulsions which were attributed at the time to the effects of cerebral birth injury were present in 7 patients during the first neonatal week, (Cases 21, 23, 37, 127, 136, 145, and 178). In Case 23, irritability was associated with the appearance of petechiae within twelve hours, jaundice within thirty-six hours and rectal bleeding within the first week. There was progressive anaemia which was corrected by two blood transfusions. In Case 127 the baby was drowsy but extremely irritable when disturbed. In Case 178 the baby could not suck for the first week and after much weight loss had to be tube fed. With the exception of Case 136, in whom there was evidence of much trauma immediately after delivery, and who had a convulsion at twelve hours, and Case 145 in which the baby seemed normal immediately after birth, all these patients had neonatal apnoea. The marked irritability and hypertonicity of the limbs settled down in four patients within two weeks, but was persistent in Cases 21, 37, and 178. In Cases 21 and 37, focal convulsions occurred within the first two months of life. Drowsiness was a more persistent symptom, however, and apathy and poverty of movement was almost universal complaints.

In /

In addition to Case 23 in which the infant became jaundiced within thirty-six hours, jaundice was noted in 6 patients within twenty-four hours. Only one of them, Case 112 had previously been apnoeic. The remaining children had appeared to be well immediately after birth, though parturition had been abnormal in four. Drowsiness and diminished tone were the commonest neurological abnormalities noted in the jaundiced patients, but in Cases 146 and 194, intermittent head retraction was observed. It is worth mentioning that both these patients were in hospital and the notes recorded the attacks though the mothers in both cases had either failed to remember or to note them. None of the patients was treated by exchange transfusion. Simple transfusions were given in Cases 23, 146, and 194. In all 7 cases the jaundice had more or less cleared by six weeks, and had been followed by pallor, commented on by the mothers in 4 cases and a persistence of their drowsiness. The jaundice in these patients will be discussed more fully later. The subsequent clinical course is considered with the neurological findings. But it should be stressed here that there was very often a latent period between the sixth week and the fourth to tenth month during which, apart from some poverty of movement, mild lethargy and some retardation of milestones, the babies do not show gross neurological abnormalities, and are often considered to be quite healthy by their parents. Only when "spasms", attacks of head retraction occur, very often with fever, or when the child begins to make voluntary movements, do they become /

become alarmed and realise that all is not well.

In addition to the six cases in which the patients showed drowsiness, reluctance to feed, and hypotonia associated with jaundice, three other patients also showed initial drowsiness (Cases 68, 173 and 202). There had been neonatal apnoea in Cases 173 and 202, but in Case 68 the immediate neonatal state had appeared to be satisfactory. The improvement in these symptoms was more gradual than that of hypertonia and excitability, and though there was some improvement in the first neonatal week, the lethargy and drowsiness persisted in most for months and was later associated with retardation of milestones, clumsiness and the onset of generalised "dystonic" spasms as in the other dyskinetic cases.

Aetiological classification of dyskinesia.

The histories of the seventeen patients which have been presented do give some basis for a reasonable classification of the disorder on an aetiological basis.

In the first place, there are the eight patients who have a history suggestive of birth injury in that parturition was disordered, and they have shown immediate neonatal abnormalities, suggesting that the abnormal parturition has resulted in damage to the child (Cases 21, 23, 37, 127, 136, 173, 178, and 202) (Table 164.).

In the second place are the two patients without evidence of clear-cut abnormality of parturition, or an immediately abnormal /

abnormal neonatal state who have, none the less, shown unusual patterns of behaviour from very soon after birth, without evidence of intercurrent disease to account for it (Cases 68 and 145) (Tables 165, 166).

In the third place are the 7 patients who have shown jaundice within 24 hours of birth, and subsequent abnormal behaviour, usually without a history of abnormal parturition, or an immediately abnormal neonatal state (Cases 23, 54, 106, 112, 146, 194 and 197).

Fourthly, there is the patient in whom there is a clear history of normal development well beyond the neonatal period and the acquisition of dyskinesia as a result of later occurring intercurrent disease, such as encephalitis, or trauma (Case 125).

These four groups of cases will be considered in more detail in order that the individual aetiological factors may be studied in more detail, and an explanation offered as to why Cases 23 and 112 appear in two categories.

Birth injury in dyskinesia.

Ten cases fell into this category. In all of them (Table 165) pregnancy and labour or delivery were abnormal as indicated, with the exception of Cases 21 and 145. In the former, the only abnormality was premature rupture of the membranes, and after a labour of sixteen hours (including a second stage of nine and a half hours) during which, apart from /

TABLE 165

Probable birth injury in patients with dyskinesia

Case No.	Parturition	Immediate neonatal state	Bth.Wgt. lbs.	oz.	Course
21	Premature rupture of membranes.	Prolonged apnoea. Asphyxia pallida.	6	8	"Cerebral, irritable, restless and hypertoxic 2 days after birth. Focal convulsions from 6 weeks.
23	Antepartum haem. Precipitate 2nd. stage. Cord round neck.	Apnoea. Cyanosed. Slow to cry. Hypospadis. Premature.	4	-	Petechiae within 12 hours. Jaundice within 36 hrs. Rectal haem. 10 days. sleepy baby. Retarded milestones.
37	Prolapse of cord.	Very prolonged apnoea.	7	2	Restless, poverty of movement. Feeble. Hypertoxic. Spasms at 4 mths. Retarded milestones.
112	Premature onset of labour at estimated 32 weeks.	Prolonged apnoea. Cyanosis.	5	9	Jaundiced from 2nd. day for 1 month. Jerking movements from 6 weeks.
127	Difficult forceps delivery.	Prolonged apnoea. Artificial respiration.	12	0	Very sleepy. But irritable and hypertoxic. "Spasms" from 10 mths.
136	1 week post mature. Difficult high forceps at 2nd. attempt.	Fractured right clavicle. Cephalhaematomata. Much bruising. No apnoea.	7	8	Generalised convulsions at 12 hrs. Jerking movements of limbs and "cyanosis" from 3 months.
173	Foetal distress during labour.	Prolonged apnoea. Cyanosed. Slow response to resuscitation. Club feet and hands.	6	8½	Sleepy baby reluctant to feed. Jerky, clumsy movements.
178	Prolonged labour. 2 weeks premature. High forceps.	Cried at once but then became apnoeic. Bruising on head.	8	12	Much respiratory difficulty. Hypertoxic. Would not suck. Restless and jumpy. Backwardness.
202	Premature labour. Prolonged 2nd. stage. 29 hours. Breech extraction. Forceps.	Brief apnoea. Cleft lip and palate.	4	4	Good progress. But sleepy hypertoxic baby with "attacks of stiffness" from 9 weeks of age.

from some maternal distress there appeared to be no abnormality. The postnatal course of the child, initially profound apnoea and asphyxia pallida, and later irritable behaviour, generalised hypertonicity of the limbs and refusal to feed, were so typical of that of the child who had suffered from cerebral damage at the time of birth, that it was decided to include the case in this category. In Case 145 labour was normal but the neonatal course was again abnormal, the child being very hypotonic and irritable. This child was therefore also included in the category of possible birth injury. None of the cases gave a history of significant abnormalities in early pregnancy apart from Case 173 in which vaccination was performed at the third month of gestation.

Only two cases gave a history of abnormality of late pregnancy, antepartum haemorrhage in Case 23, and in this patient there were other possible causes of cerebral damage since the second stage of labour was precipitate and the cord was round the neck when the child was delivered. In Case 68 there was antepartum haemorrhage and a prolonged first stage of labour.

The remaining abnormalities were confined to labour and delivery and consisted of prolapse of the cord in Case 37, foetal distress in Case 173, and difficult instrumental delivery for a variety of indications in Cases 127, 136, 178 and 202.

Seven of the patients in this group were apnoeic for a period after delivery. Case 136 showed cephalhaematomata and /

Table 166.

Possible birth injury in patients with dyskinesia.

Case No.	Preg. Labour and Delivery.	Immed. Neonatal State	Birth weight. pounds/ounces.	Course.
68	Antepartum haem. after fall. Prolonged Ist stage.	Normal	6/13	Very placid lazy baby. Clumsy from 6 months. Unsteady from one year.
145	Labour of 2 hours.	Normal.	7/2	Irritable very hypotonic baby.

and much skull bruising, together with a fractured clavicle indicated that some trauma had probably resulted from the repeated attempts at forceps delivery. Cases 68 and 145 were normal. In Case 23 jaundice developed within 36 hours and the child was drowsy and apathetic. There was, in addition, petechiae and rectal haemorrhage. Of the remaining cases, three were noted to be very drowsy for some days after birth and to be reluctant to feed (Cases 68, 173 and 202), and six showed the picture of gradually diminishing irritability, hypertonicity of the limbs and failure to feed. These symptoms were associated with convulsions at 12 hours in Case 136, and by intermittent twitching within a few days in Case 37. In Case 21, focal convulsions ensued within six weeks.

In attempting to generalise about these cases in whom the postnatal course was so suggestive of birth injury, the most that can be stated is that the abnormalities of parturition occurred almost exclusively during labour and delivery and that they appeared to be as often traumatic as hypoxic. Multiple disorders of pregnancy, labour and delivery appear to be much less frequent than, for example, in congenital hemiplegia, but to be attended by prolonged apnoea and later "cerebral" behaviour in a rather higher proportion of cases.

The relationship between the abnormalities of parturition and the neonatal course in Table 165 seems to be in favour of birth injury in the cases in Table 166. Birth injury cannot be excluded in these two cases [redacted], though the

evidence /

TABLE 167

Patients suffering from dyskinesia with a history of prolonged neonatal jaundice

Case No.	No. of Preg.	Parturition	Haematology	Bth.Wgt. lbs.	oz.	Neonatal state	Neonatal course	Subsequent findings
23	1st. and only.	Slight antepartum haem. Cord round neck.	Mother Rh+ve Child Rh+ve Coombs test +ve	4	-	Apnoea. Hypospadis.	Petechiae in 12 hrs. Jaundice in 36 hrs. for 2 wks. Rectal haem. 1 week.	Sleepy baby. Retarded milestones. Later mild choreathetosis.
54	5th.of 5	Normal.	Mother Rh-ve Child Rh+ve(1952) No persisting maternal anti-bodies.	6	4	Normal. Cried at once.	Jaundiced within 6 hrs. for 3 wks. Then pale, hypertoxic and irritable.	Mild generalised chorea.athetosis. Restricted eye movements.
106	2nd. of 2	Rather rapid labour of 2 hours.	Mother Rh-ve Child Rh+ve.No persistent anti-bodies.(1953)	8	8	Normal.	Jaundiced within 12 hrs. Oedematous. Drowsy. Hypotonic.	Mild generalised choreathetosis. Restricted upward gaze.
112	1st.and only	Premature onset of labour 32 weeks.	Mother Rh-ve. Child Rh+ve No persistent antibodies(1953)	5	9	Prolonged apnoea.	Jaundiced from 3 days for 1 month.	Generalised chorea-athetosis.High frequency deafness. Limitation of upward eye movements.
144	2nd. of 2	Post mature 43 wks.	Mother Rh-ve Child Rh+ve Persistent anti-bodies (1953)	7	8	Normal.	Jaundiced within 24 hrs. for 4 wks. Blood transf. Floppy.	Mod.severe chorea-athetosis.High tone deafness. Restricted upward gaze.
194	2nd. of 2	Chronic nephritis and pre-eclampsia.	Mother Rh-ve Child Rh+ve Coombs positive.	8	3½	Normal.	Jaundiced within 24 hrs. (Hb 80%), for 6 weeks. Hypotonic. Broncho-pneumonia.	Jerking attacks from 4 mths. Generalised choreathetosis. Eye movements restricted.
197	3rd. of 3	Precipitate.Premature labour.Spontaneous delivery.	Mother Rh unknown Paternal Rh+ve	4	4	Jaundiced. Breathed at once.	Progressive jaundice for 5 days - persisted 6 wks. Drowsy and irritable until 12 weeks of age.	Generalised chorea-athetosis.Paresis of upward gaze. Hearing doubtful.

evidence for it is much less convincing. It seems probable, however, that abnormal processes in labour or delivery, whether liable to cause hypoxia or trauma when associated with post-natal apnoea are relatively frequent antecedents to dyskinesia, and that this combination may be recognised in approximately half the patients in this series.

Cases with a history of jaundice in the neonatal period due to blood group incompatability.

Seven cases fall into this category, including Case 23 which has already been described (Table 167). All of them developed jaundice within thirty-six hours, all most within twenty-four hours, and they remained jaundiced from 2 weeks (Case 23) to 6 weeks (Cases 146 and 197). Their clinical findings in the neonatal period have been described briefly. None of them was exchange transfused. Haematological tests were performed at the time of pregnancy, or following the appearance of jaundice in only two of the patients. Three were born before facilities for testing were available locally (about 1947), and in two there was a failure to use the facilities which were available by the time the children were born. In Case 23, tests for rhesus incompatability were performed when the child became jaundiced during the second day of life. These showed that both the child and the mother were rhesus positive and the Coombs' test was weakly positive. No tests for ABO incompatability were performed (1948). Testing in 1953 confirmed the rhesus grouping and since the patient was the first child ABO

incompatability /

incompatibility remained a possibility.

Of the remaining patients one was shown to be suffering from the effects of rhesus incompatibility when she became jaundiced, though during the mother's pregnancy no antibodies were detected on repeated examinations (Case 194). In a postnatal specimen of the mother's blood antibodies were found 1/4 in saline and 1/6 in albumin, the child's Coombs' test being positive.

In the remaining cases retrospective diagnosis had to be made on the basis of the history of jaundice and the other clinical evidence, together with the results of grouping blood obtained at the time of the survey. All except Case 197, in which the mother refused the needle, child and mother were found to be rhesus incompatible, and that in one case, weak maternal antibodies were detected as long as five years after the time of delivery, a rather unexpected confirmatory piece of evidence. Unfortunately the blood of the fathers was not obtained. With the exception of Case 23, therefore, it seems probable that the jaundice from which the patients suffered was due to rhesus incompatibility. The descriptions of the neonatal behaviour of the seven patients strongly suggests that they suffered from kernicterus. Additional evidence for this conclusion is obtained from the results of later neurological examination, for it has been shown that impairment of conjugate eye movements, especially convergence and upward gaze, and high tone deafness occur much more frequently in dyskinetic patients after kernicterus than when brain /

brain damage is due to other causes (Byers et al, 1944).

The only patient of the seven with neonatal jaundice who did not show impairment of upward gaze was Case 23, and in at least two patients the suspicion that there was high tone deafness was confirmed by audiometry (Cases 112 and 146). In Case 197 hearing seemed to be defective but the patient was untestable.by audiometry.

Whilst blood group incompatibility appears to have been present in all the patients, with the possible but unlikely exception of Case 23, the proportion with indications that hypoxic or traumatic brain damage may also have occurred is noteworthy. Pregnancy, labour or delivery were abnormal in Cases 23, 146, 194 and 197, and whilst they appeared uncomplicated in Case 112 the child was born in a state of asphyxia pallida and suffered from prolonged apnoea. Cases 112 and 197 were premature by weight. In Cases 23 and 112 especially the immediate postnatal state was strongly suggestive of severe birth injury.

These findings obviously suggest that cerebral hypoxia or trauma may have been of contributory aetiological importance in causing brain damage in a proportion of these patients who were jaundiced as a result of blood group incompatibility. This is in accordance with the findings of other workers who have described what appears to be an increased tendency for premature or hypoxic brains to retain circulating bilirubin, especially in the brain stem and basal ganglia (Aiden et al, 1950).

Since /

Since it is impossible to determine the relative importance of kernicterus and of paranatal hypoxia and trauma in Cases 23 and 112 they have been included in both the descriptive categories of "Birth Injury" and "Neonatal Jaundice". It seems clear, however, that blood group incompatibility and cerebral hypoxia or trauma may be contributory and very closely related in a high proportion of patients suffering from dyskinesia.

CHAPTER 11 c

The clinical findings in seventeen patients suffering from dyskinesia.

The development of dyskinesia. The fully developed clinical picture of dyskinesia is not usually manifest until after the second year of life, or even later. The characteristic involuntary movements found in the condition appear gradually many months after birth and are not found in the newborn child.

As in diplegia, however, various arbitrarily defined stages in the course of the development of the final clinical picture may be recognised, whether the dyskinesia is the result of "birth injury" or "kernicterus". These stages are characterised by clinical findings which allow a fairly confident diagnosis of dyskinesia to be made in the majority of patients considerably before the final clinical picture emerges.

The arbitrarily defined stages observed in the majority of the 17 patients in this series may be described in more detail. In the newborn period most patients suffering from dyskinesia show rather non-specific abnormalities of behaviour indicating the probability of brain damage, either due to "birth injury" or "kernicterus", rather than suggesting that the child will later suffer from dyskinesia. These non-specific abnormalities of behaviour may persist for days or occasionally two or three weeks, but as the child matures they gradually disappear. There follows a stage in which abnormalities of behaviour are much less striking though retardation of motor development and abnormalities of muscle tone /

tone may be demonstrated by examination. This may be termed the latent or hypotonic stage. It lasts for between four and ten months in most cases. Involuntary movements appear only at the end of this time. They are commonly similar to those observed in diplegia, comprising generalised extensor movements of the trunk and limbs, especially when the child's position is changed or the head is allowed to extend. They may be termed "dystonic". Over a period of many months the involuntary movements gradually become less generalised, and instead of involving a mass movement of the trunk and all four limbs, more discrete patterns of involuntary movements affecting individual limbs emerge. The final picture of dyskinesia may be recognised, with characteristic findings of choreoid, athetoid, or dystonic movements and of tension in the limbs. As in diplegia the most severely affected patients will show features of the earlier stages of dyskinesia. Very badly damaged patients may be immobile, hypotonic and extremely helpless for the rest of their lives, and in a larger proportion of less severely affected dyskinetic children fragments of the mass movements characteristic of the dystonic stage may be noted. The most fortunate children suffering from dyskinesia lose most of the typical manifestations of the earlier stages. Their involuntary movements affect discrete parts of the limbs only, and voluntary activities are accordingly much less impaired.

The neonatal period. The abnormalities of the immediate and later neonatal period shown by patients were fully described /

described when the aetiology of dyskinesia was considered. All the cases thought to be congenital showed abnormalities in the neonatal period suggestive of disorder of the central nervous system.

Six appeared to be normal immediately after birth. Eight were apnoeic, one after taking a few initial breaths. One (Case 197) was jaundiced and one (Case 136) showed evidence of widespread trauma, but both these patients cried at once. Congenital malformations were observed in Cases 23 and 202.

Severe jaundice developed in seven patients. Six of these were described as excessively drowsy and lethargic (Cases 54, 106, 112, 146, 194 and 197). Three were recorded in hospital notes as having a tendency to head retraction during the first two to three weeks. Hypotonia was observed in four. Irregular jerking of the trunk and limbs especially when being handled was noticed by the mother when the patient was one month of age in Case 54. One patient with a hypospadias had numerous petechiae for a week after delivery and a rectal bleed on the seventh day requiring blood transfusion. The clinical picture in this patient was rather different. He is recorded as being restless and irritable, with jerky movements of the limbs and a tendency to spontaneous head retraction. He remained irritable for between one week and ten days and then became more like the others in behaviour, showing lethargy and generalised hypotonia (Case 23). The clinical picture in these seven patients is compatible with their /

their having suffered from kernicterus, though the clinical details are less full than one would have wished. By the time the jaundice had faded, usually before the age of 6 weeks, the abnormalities of behaviour had become much less marked. Drowsiness, lethargy or irritability had decreased but all the patients were noted to be rather hypotonic and less active and alert than normal.

Of the nine patients who were not jaundiced, six were noted to be excessively irritable, hypertonic and "jumpy" shortly after birth (Cases 21, 37, 127, 136, 145 and 178). Case 136 had a generalised convulsion at 12 hours. Fixing to the breast was usually difficult and feeding was slow. In three patients the hypertonicity and irritability became less marked within two weeks but lasted for about three weeks in Cases 21, 37 and 178. All these patients then showed a picture of poverty of spontaneous movement, hypotonia and apparent lethargy.

The other three patients who were not jaundiced showed persistent lethargy and drowsiness from shortly after birth Cases 68, 173 and 202), which persisted in all for three to four weeks and showed only gradual improvement after this time. Slow feeding was troublesome in Cases 68 and 173 because there was a tendency for the babies to go to sleep during feeds. But the neonatal abnormalities in these patients were less well defined and striking than in the drowsy jaundiced babies or the restless irritable patients.

It is apparent from these findings that a variety of abnormalities of behaviour in the neonatal period occur in dyskinetic /

dyskinetic patients. Though the clinical picture of kernicterus immediately gives rise to the suspicion that the child may later show dyskinesia. The actual abnormalities of behaviour encountered in the neonatal period are not characteristic of dyskinesia. The same drowsiness, irritability, restlessness, twitching and tendency to head retraction may be observed in babies who later show other forms of cerebral palsy or mental defect, or who are later apparently quite normal.

When the initial abnormalities of behaviour have disappeared the infant is usually found to be somewhat lethargic, inactive and hypotonic whatever the type of his neonatal behaviour disorder. Thereafter abnormalities of development may be noted which are rather characteristic of dyskinesia.

The hypotonic (or latent) stage. For a period after the patients had ceased to show the classical manifestations of kernicterus or "birth injury" their behaviour was often considered to be normal by their parents. In retrospect many of them described some lethargy, lack of interest in the environment, poverty of movement, and hypotonia. Compared to older siblings they were slower in smiling, following objects with the eyes, and holding up their heads, but the children were not conspicuously abnormal. They were often described as being particularly "good" babies. When this stage persisted beyond five or six months, feeding difficulties were liable to appear. In ten of the sixteen congenital cases the children found great difficulty in taking semi-solids from a spoon and were described as holding food in their mouths, or pushing /

pushing it out with their tongues instead of swallowing. At about the same time dribbling of saliva from the mouth often became a problem. One patient began to suffer from and continued to have epileptic convulsions during the hypotonic stage (Case 21).

On examination in the hypotonic stage the striking findings were the retardation of neuromuscular development, poverty of movement, and generalised hypotonia. The babies were usually thriving quite well and were within the average range for weight, height and head circumference.

Apart from pallor, observed especially in anaemic post-kernicteric patients, the general appearance was not one of ill-health.

Developmental milestones had not been reached at the normal times, however. Smiling was not usually much delayed but the ability to focus with the eyes on objects in the surroundings and to follow them was achieved late. Four patients were noted by paediatricians to have apparent difficulty in focussing on objects, though blindness was seriously suspected in only one (Case 194). Parents suspected blindness more frequently, but hesitated to mention their fear in case it might be confirmed by the doctor. Even when the child had learnt to follow objects, difficulties in fixation of the eyes persisted and involuntary movements of the eyes were frequently noted. Upward gaze was restricted in all the post-kernicteric patients even at the time of examination during the survey.

Impairment of hearing was suspected in four post-kernicteric patients before they were six months of age, since they did /

did not react to noises in their immediate environment. Three were later found to have significant high frequency deafness. Patients over the age of four or five months, especially when they were teething, were noted to be salivating profusely and not to be swallowing saliva as frequently as normal babies.

Control of the head was consistently poor, the youngest age at which it was achieved by patients in the post-kernicteric group being 5 months (Case 106). When picked up, the babies felt "floppy" and parents often described their children as being "like a bag of beans". Poverty of movement was striking. Bed clothes tended not to be disturbed for hours at a time, and the normal tendency for babies of 4 to 6 months to reach for objects was not noted until very much later.

Reflexes characteristic of the neonatal period can usually be elicited for a much longer period than in the normal child. Thus Moro responses, symmetrical and asymmetrical neck reflexes, grasp reflexes, and sucking reflexes are brisk and extremely easy to elicit in the majority of patients in the hypotonic stage of dyskinesia. Unfortunately records of examinations performed on the patients in this series rarely mention them. Tone in the limbs was noted to be reduced in 11 of the 16 congenital cases when the patients were at rest. Tendon jerks were not often recorded but when they were they tended to be sluggish, which is in accordance with later personal experience of other patients (Yannet and Horton, 1952).

The /

Figure 13 . Severe dyskinesia in girl of average intelligence. a.Symmetrical neck reflex.b.Tonic neck reflex.c.Moro reflex.

a.

b.

c.

The duration of the hypotonic stage was variable. It came to an end rather dramatically in 9 of the 16 patients suffering from congenital dyskinesia with the appearance of sudden mass involuntary movements involving extension of the trunk and all four limbs, when the child was startled or handled, especially if the head was allowed to extend. The appearance of these mass dystonic movements usually coincided with the child gaining some head control. In three they first occurred when the children were febrile as a result of respiratory infections. The patient in Case 194 was under close personal observation at the time of the first appearance of these movements. When the respiratory infection cleared up so did the involuntary movements, only to return with another respiratory infection. Thereafter they were persistent.

In the other 7 patients suffering from congenital dyskinesia no history of the relatively sudden appearance of mass involuntary dystonic movements was elicited. In them the end of the hypotonic stage was marked by the appearance of slow irregular involuntary movements predominantly in the limbs whenever the child attempted voluntary movement or when his position was changed passively. In two patients the child's upper limbs movements were described as "grossly clumsy and unsteady" and as "grossly ataxic and wild" respectively. It is characteristic of patients showing these early involuntary movements in the upper limbs that there are involuntary reciprocal pedalling movements in the lower limbs when the child is held upright. These were

not /

Case 178.

Very severe dyskinesia in a boy whose stage of generalised hypotonia and poverty of movement was prolonged, and onset of dystonia was not abrupt.

He was the only child of healthy parents, the mother being 38 at the time of his delivery. The pregnancy was uncomplicated apart from morning sickness during the first three months. Labour occurred spontaneously at an estimated 38 weeks' gestation after the spontaneous rupture of membranes. After 48 hours of dry labour a high forceps delivery was performed with great difficulty, and only after manipulations lasting for two hours with the mother under chloroform anaesthesia. The birth weight was 8 lb. 12 oz. and the child cried at once but thereafter stopped breathing and required oxygen and resuscitation. For the first week after delivery the child lay with the hands clenched tightly, and he seemed stiff and was irritable when handled. He had to be fed from a spoon or tube for 3 weeks as he could not suck. Bruising over the head and occiput disappeared gradually during this time. Large amounts of phenobarbitone and chloral were given in an attempt to improve his irritability. He contined to be irritable when disturbed and cried a great deal at night, but never made much spontaneous movement during his first year. After the first 3 - 4 weeks the position of generalised flexion associated with stiffness of the limbs was replaced by a generalised "floppiness" though the fingers continued to be clenched over the adducted thumbs. When picked up "his head lolled about" and it continued to do so until the age of about 18 months when he began to achieve some head control. He was able to sit with support by the age of two years, crept at three and smiled at the age of about one year.

He began to reach for objects at the age of about two years, and when he did so it was observed that he was very clumsy and inaccurate, flail-like motions of the upper limbs resulting when he tried to direct them voluntarily. The upper limbs used to go stiff in extension, often knocking the desired object out of reach, and there were associated involuntary movements of extension in the trunk, head and neck. "Pedalling motions" of the lower limbs were also apparent at these times. At rest he continued to feel floppy, but less so than previously and his head control gradually improved. When he was handled or alarmed, however, he would suddenly go stiff, with the upper limbs extended and the head and trunk extended and rigid. When he was sat up and became in the least excited or active the stiffening in extension of the trunk and upper limbs might be very violent and his lower limbs had to be restrained because the involuntary reciprocal pedalling movement they showed caused skin abrasions.

The involuntary movements of trunk and limbs whenever he attempted voluntary motion gradually diminished in severity as /

as he grew older, and by the age of 7 he was able to take up to twenty steps in light calipers with support on either side and was able to reach for large objects with the left hand with fair success. His first words had been spoken at the age of 4 years, but he was unable to say more than two-word phrases and these were unintelligible to all but his parents until he was 7 to 8 years of age, on account of the severe involuntary movements of his tongue and "his difficulty in getting breath to speak with". He drooled continuously.

From the age of seven his findings changed little. He attended a school for spastic children as a day pupil and was thought by his teachers to have learnt to read enough to make reading pleasurable by the age of 8 years.

On examination aged 12 he was a rather pathetic figure, mouth breathing, salivating profusely, unable to sit without support or stand, with poor head control, and could make only very rudimentary articulatory efforts. He comprehended most of what was said to him, as far as could be determined, obeyed quite complicated commands, and had a sense of humour and at least some imagination. He could see quite small objects, but his visual fixation was very poor, so poor in fact that I was surprised that he was able to follow a printed line. His fundi were normal; eye movements were full and quick, though rather irregular and at times accompanied by nystagmus.

He could shut his eyes and open them to command; facial sensation was normal; his face was expressionless; involuntary chewing movements of the jaws occurred constantly and were associated with irregular protrusion and retraction of the tongue and swallowing. He could hear even a whisper. Tongue movements were very poorly controlled and he could not maintain voluntary protrusion of the tongue for any length of time. The power in all four limbs is diminished so far as voluntary movement is concerned. Power was more severely impaired in the distal parts of the limbs than in the proximal, and in the right upper limb than the left upper limb. Head control was extremely poor, and he appeared to have virtually no voluntary sitting control of the trunk muscles. More impaired still than the power of voluntary movement was his co-ordination. He could not perform the finger-nose test with either hand, though he attempted this, and he found even voluntary flexion of the fingers difficult to achieve on command. All attempts at voluntary movements were frustrated by intense choreoid and dystonic movements. The latter were more prominent on the left side than on the right, and the right arm tended persistently to go into an opisthotonic position. This tendency was less marked on the left side though it was also present there. Choreoid movements were more marked in the left upper limb than on the right. In both hands athetoid movements were also present. Much of his motor activity was still dominated by his tonic neck reflexes. These were very marked, and any change in his /

his position resulted in a turn of the head, and subsequently in very rapid alterations of muscle tone and position in all four limbs. Tone in the limbs when he was asleep was relaxed, but at rest there was rigidity in the right upper limb of very marked degree, and this was also present in the left arm. In the left arm and in both legs, however, there was considerable spasticity, and in these limbs the biceps, triceps, supinator, knee and ankle jerks were grossly increased. The plantar responses were extensor. I could not test sensory findings fully, but he certainly appreciated light touch, pin-prick and vibration in all four limbs.

During the examination there were several bouts of very active chewing and swallowing and tongue movements, sometimes associated with slight jerking movements of the head and a very definite generalised increase of tone. He seemed to have some impairment of consciousness for 3 or 4 seconds, during which these movements were present. His level of consciousness appeared to fluctuate rather from minute to minute, which did not make assessment of his awareness of things any easier.

The psychologist was in little doubt that he was well below normal on all tests of intelligence which were possible, but the fluctuations in his performance from hour to hour, from day to day, and from month to month, made it very difficult to say much more than this. In general, however, he felt that nothing the boy did was above an eight-year level.

not noted to have occurred by those who examined any of the 7 patients in the series during infancy. Three showed these pedalling motions at the time of examination during the survey, however, and their parents stated that the children had shown them from the early months of live. (Cases, 136, 178 & 202). Though mass dystonic movements may be demonstrated in patients whose discrete involuntary movements have followed directly upon the hypotonic stage, the majority do not appear to show the same period during which intense powerful mass extensor movements are the presenting abnormality, called the dystonic stage. They tend to go straight into the stage of discrete involuntary movements.

The hypotonic stage persisted until between 3 and 20 months after birth in the 16 patients suffering from congenital dyskinesia. In the majority it was replaced by the dystonic stage, or the stage of discrete involuntary movements between the ages of 4 and 10 months. Patients with the most prolonged hypotonic stage were generally those who were later found to be most handicapped, for example, Case 178.

The Dystonic Stage. The involuntary movements first shown by the dyskinetic child when he emerges from the hypotonic stage tend to be generalised, affecting the trunk as well as the limbs to a greater or lesser extent. They resemble the "dystonic attacks" shown by diplegic patients very closely. Alarm, sudden changes of position, and especially extension of the head result in a generalised involuntary /

involuntary movement in which the head and trunk become extended and the limbs assume a "decerebrate position". This consists in the upper limbs of adduction at the shoulders, extension of the elbows, pronation of the forearms and flexion of the wrists and fingers, the thumbs usually being adducted in the palms. The lower limbs are extended and adducted at the hips, extended at the knees, with the feet in plantar flexion and inverted. There is a great increase of anti-gravity tonus and the body and limbs feel rigid while the position is maintained, usually for only a few seconds.

Parents note that their children tend to "back arch" when being bathed or dressed. When placed with support in the sitting position, recurrent back arching pushes the child's bottom forwards, and in a series of jerks he again reaches the supine state. A loud noise, such as the banging of a door, or a nearby factory siren will cause the position to be assumed to the distress of the child who may cry out as the involuntary movement occurs.

Nine of the 16 patients suffering from congenital dyskinesia suffered from such attacks which occurred after a period during which their parents had assumed they were making satisfactory progress in spite of their retarded motor milestones. Their onset in four was so dramatic that the patients were taken for medical advice within a few days of their appearing. In the other five the attacks were relatively mild for a period of a few weeks, but gradually became more readily precipitated and more severe.

On /

On examination during the period when involuntary generalised dystonic attacks are occurring frequently the patients show a remarkably consistent stage of neuromuscular maturation. They can hold their heads up when placed in the erect position reasonably steadily, but cannot sit with support, reach for objects successfully, or bring objects placed in their hands to the mouth. The Moro reflex is still present, though if the head is allowed to extend the response to alarm is usually to have a generalised dystonic movement rather than a Moro response. The symmetrical neck reflexes are extremely brisk and brisk extension of the head immediately results in a generalised dystonic movement. The tonic neck reflexes are marked. The grasp reflexes are present. The sucking reflex is usually very easily elicited.

As the child matures the dystonic movements become less troublesome, and the range of his voluntary movements becomes wider. Whereas at the beginning of the dystonic stage any attempt at voluntary movement promptly resulted in a generalised dystonic movement this is no longer the case. He may manage to get the hand to mouth, roll over from the supine position and reach for objects more successfully. Head control improves and sitting with support for a longer period becomes possible. The dystonic movement becomes less generalised when it does occur. Instead of a generalised involuntary movement of the trunk as well as all the limbs, the less affected limb, commonly an arm, is relatively spared and may continue to show a position of semiflexion during the attack. /

attack. To produce a generalised attack at this stage of neuromuscular maturation, further stimuli to the reflex increase of antigravity hypertonus are required. Thus, placing the child in the erect position with pressure on the soles of his feet will usually result in a generalised dystonic movement, and an increase in antigravity tonus. Reflex standing may be induced in a high proportion of dyskinetic patients by these manoeuvres.

On examination during the dystonic stage the children are found to be very helpless. Though some independent head control has usually been gained, even supported sitting is impossible. Hand use is very limited and reaching for objects is attempted in a very tentative and primitive fashion. When quite at rest there is usually generalised hyptonia with rather sluggish biceps, triceps supinator knee and ankle jerks, the plantar responses being infantile. Sucking grasp, symmetrical and tonic neck reflexes are very marked. The Moro reflex is present. Any jerky passive movement or attempt at voluntary movement is likely to result in the occurrence of a generalised mass involuntary movement, the child assuming the generalised position of extension in the trunk and all four limbs which has been described.

As the generalised dystonic movement becomes less generalised and less readily elicited the dystonic stage may be said to merge into the final stage of dyskinesia. A greater range of voluntary activity becomes possible and the involuntary movements which are produced when voluntary movement is attempted /

Case 194.

The onset of the dystonic stage of dyskinesia personally observed in a girl who suffered from kernicterus.

This child was the result of the second pregnancy to a mother aged 25 who was rhesus negative and said to suffer from "subacute nephritis". Her husband and first child were rhesus positive. During the pregnancy she had three attacks of haematuria and hypertension, one of which, at the fifth month of gestation, was severe enough to necessitate hospital admission. No antibodies were detected in repeated blood samples.

The child was delivered in hospital spontaneously at term after an uncomplicated labour of 2¾ hours, with a birth weight of 8 lb. 3¼ oz. She breathed at once. Twenty-four hours after birth she was noted to be very jaundiced. The Coombs' test was positive and the haemoglobin 80%. She was rhesus positive. She remained jaundiced for five weeks and required five blood transfusions during this time in order to maintain her haemoglobin. For four weeks her spleen was palpable. She was tense, irritable and restless from the second to the twelfth day of life and showed intermittent irregular twitching movements of the limbs. There was a tendency to retract her head which was noted by the mother during feeding. She was discharged from hospital at the age of eight weeks and was seen frequently in the Infant Clinic of the hospital, though no neurological abnormalities were noted at the time of discharge.

By the age of 18 weeks she was smiling, looking about her though she could not look upwards, holding her head fairly steadily, and was beginning to sit with the support of two pillows. At this time she developed an upper respiratory infection with cough, mild fever and seemed rather fretful and miserable. There was poverty of movement of the limbs and when she did move them she did so jerkily and incoordinately - "kind of throwing them, as if she didn't know what to do with them", said her mother. Whenever she tried to move her trunk or her position was altered passively, sudden extension of the head, neck and trunk occurred with a marked generalised increase of tone, the limbs assuming extended positions as in opisthotonos. Her mother observed that she was unable to sit as she had been doing previously, and said that on handling her "it felt as if she was like a spring; suddenly she goes backwards as stiff as a board." The position of generalised extension was maintained for only a few seconds and there did not appear to be any impairment of consciousness during her "extensor attacks". She seemed normal after them. They continued to occur whenever she was handled for five days, but as her cough and fever improved they became less frequent and severe and at the end of ten days had disappeared.

On /

On examination 14 days after the onset of her attacks she was afebrile and well. She was again able to sit with support, and her limb movements were within normal limits for her age.

At the age of 20 weeks, however, there was a recurrence of cough and fever, and the attacks recurred whenever she was handled or her position was changed, or when she tried to move herself. They were identical to those observed previously. Though her fever and cough disappeared within ten days, the involuntary extensor movements persisted. She was unable to sit or reach for objects as previously, and seemed less interested in her surroundings. The attacks gradually diminished in frequency and severity between 26 and 28 weeks and finally ceased to occur when she was aged 30 weeks.

At this time, though generalised involuntary movements no longer occurred, it was noted that whenever she tried to move either upper limb a sudden movement of adduction and slight flexion of the shoulder, extension of the elbow, pronation of the forearm and flexion of the wrist and fingers over the adducted thumb occurred. Attempts to move the legs resulted in extension and adduction at the hips, extension of the knees, plantar flexion of the feet. In these positions the limbs felt very stiff. She was unable to hold the head steadily when her position was changed. She drooled from the mouth.

She began to sit with support again at the age of 8 months and was reaching for objects successfully by this time. Involuntary movements of the upper limbs were less severe, and she appeared to be able to resist the tendency for them to be involuntarily extended more successfully.

At the age of 13 months she was alert and interested in her surroundings, and was noted to have seven greenish teeth which were fragmenting. There was a convergent squint and bilateral nystagmus on lateral gaze, and she would not look up. There was a facial asymmetry, the face being pulled slightly to the left. Her hearing appeared to be impaired. Her right upper limb was used stiffly in the extended position to reach for objects, and she appeared to have little control over the movements of the fingers. The left upper limb was used very little and tended to be held by the side with the shoulder adducted, the elbow extended, the forearm pronated, and the fingers flexed over the adducted thumb. All movements with either upper limb were very jerky and irregular. Any attempt at voluntary movement resulted in a marked rigid increase of tone most evident in the parts she was moving but affecting all parts of the body.

At rest there was some generalised increase of tone in all four limbs with some exaggeration of the biceps, triceps, supinator, knee and ankle jerks bilaterally, more on the left than the right. The abdominal reflexes were present, and the plantar responses were infantile in type. Sensory findings were untestable.

attempted tend increasingly to be confined to discrete parts of the body, and less generalised. Thus, when the child attempts to reach for objects the operating arm is liable to show quite gross involuntary movements and a tendency to assume a position of adduction at the shoulder, extension of the elbow, pronation of the forearm and flexion of the fingers and wrist, accompanied by an increase of antigravity tonus, but the trunk will not be involved to the extent that it should have been a few months before. Generalised dystonia is gradually replaced by involuntary movements of discrete parts of the body.

Because the manifestations of dyskinesia only gradually cease to be generalised over a period of months it is difficult to state arbitrary criteria by which the end of the dystonic stage may be judged. Achieving the ability to sit with support and to put an object placed in the hand to the mouth without there being a generalised dystonic movement are probably the best indications that the dystonic stage is ending. Using these criteria, the dystonic stage lasted until the age of 9 months in 3 patients, 18 months in 2, over $2\frac{1}{2}$ years in 3 and in 1 at the age of 7 (Case 194). At least four of the other seven patients suffering from congenital dyskinesia in whom the relatively sudden onset of generalised involuntary movements had not been observed, dystonia was certainly present for a period. Unfortunately it is impossible to obtain any accurate idea as to how long generalised dystonic attacks persisted in these cases. They

were /

were probably present until the age of between 2½ and 3 years in 2 of them.

The stage of discrete involuntary movements. The most severely affected patients suffering from dyskinesia continue to show the features of the hypotonic stage, the slightly less severely affected patients the rather more mature neuromuscular patterns characteristic of the dystonic stage. In the majority more localised involuntary movements involving the trunk and all 4 limbs. Commonly residual fragments of the movement patterns typical of the dystonic state persist into the stage of discrete involuntary movements. Reflex stimuli which increase antigravity tonus, such as placing the child in the erect position with pressure on the soles of the feet and extending the head cause an extensor movement of the trunk and the lower limbs, though the arms take less part in the movement than they do in the dystonic stage. In some patients this "extensor thrust" (as it is known to therapists) is a major factor in preventing unsupported walking, though in less severely affected children the tendency to back arching in the erect position is more of clinical interest than it is an actual disability. In the upper limbs fragments of the mass dystonic movement characteristic of the previous stage may also be recognised. Voluntary movements of the upper limbs tend to produce involuntary movements which result in a position of adduction at the shoulder, extension of the elbow, pronation of the forearm and flexion of the wrist and fingers - the "decerebrate arm".

Patients /

Patients who do not pass through the dystonic stage but proceed straight from the stage of hypotonia to the stage of discrete involuntary movement show much less tendency to antigravity hypertonus in either the upper or lower limbs during the stage of discrete involuntary movement. In them the major causes of failure to walk are the persistent reciprocal treading movements in the lower limbs and the poor postural control of head and trunk.

Whether as a result of involuntary antigravity hypertonue associated with extension of the trunk and lower limbs, or of reciprocal involuntary treading movements, the majority of patients suffering from dyskinesia walk late. Only 2 were able to walk unsupported before the age of 18 months amongst the 16 children suffering from congenital dyskinesia. (Cases 54 and 68). Six others walked at or before the age of 3 years, 5 between the ages of 3 and 6 years, and 3 had not achieved independent walking by the age of 7. (Cases 136, 178 and 194).

The findings in the patients in the stage of discrete involuntary movements will now be considered in more detail.

The types of involuntary movements. Five types of involuntary movements were recognised though transitional types of movement occurred not infrequently which were difficult to classify. The majority of patients showed 2 or more types of involuntary movement.

Athetoid /

Athetoid movements were similar to those observed in hemiplegia. They consisted of slow writhing movements of the distal parts of the limbs, affecting most severely the fingers, wrists and forearms. The affected hand most commonly showed slow extension and abduction of the digits, extension of the wrists and supination of the forearm, followed by flexion and adduction of the digits, flexion of the wrist and pronation of the forearm.

Choreoid movements were similar to the involuntary movements found in rheumatic chorea, though their speed tended to be rather slower and they affected larger portions of the anatomy in most cases. In contrast to athetoid movements, choreoid movements were more marked in the proximal parts of the limbs than the distal and were relatively rapid and jerky. They were organised movements in the sense that they involved reciprocal action of synergist muscles and relaxation of antagonists. They were commonly more marked on one side of the body than the other, and frequently affected the muscles of the face and tongue in addition to those of the limbs.

By tension was understood a sudden increase of muscle tone in the limbs, usually more marked in antigravity muscles than in flexors. The increase of tone was usually sudden and very marked, sufficiently powerful in many patients to arrest all movement of the affected part. It was most commonly precipitated by attempts at voluntary movement and affected the moving part predominantly though it commonly involved other limbs /

limbs and parts of the trunk also. Sudden generalised muscular hypertonus, or tension, was also commonly produced by alarm or sudden passive movement. Tension occurred invariably in choreoid and dystonic movements and to a lesser extent in the course of athetosis. In the sense that it more frequently caused an arrest of voluntary movement than a definable involuntary movement, the inclusion of tension in a discussion of involuntary movements may seem illogical. But tension is, in fact, a manifestation of the same incoordinate uninhibited nervous activity as causes involuntary movement, (Jakob, 1925).

By dystonia was understood a relatively slow involuntary movement with trunk involvement in which there was a marked increase of antigravity tonus and a tendency for the affected part to assume the position found after a mass dystonic movement. In a number of cases the description of dystonia given by some recent authors of "slow chorea" was appropriate. Dystonic movements may be regarded as representing fragments of the mass dystonic movements of the dystonic stage of dyskinesia. They were never the only form of involuntary movements in the patients in the present series, but always accompanied by either choroid movements or by athetosis. Tension was invariably present, and indeed comprises part of the picture of a dystonic movement.

The functional impairment of voluntary movement in cases of dyskinesia. It is a matter of the greatest difficulty to detect to what degree the impairment of voluntary /

Table I68.

The types and distribution of involuntary movements in I7 patients with dyskinesia.

Distribution of movements.	Hemiplegic.	Triplegic.	Tetraplegic.	Totals.
Athetosis alone.	0	I	0	I
Choreoid only.	0	0	I	I
Athetoid and choreoid only.	0	4	2	6
Athetosis with tremor.	0	0	I	I
Choreoid movements, athetosis and tremor.	0	0	I	I
Choreoid movements with dystonia.	0	0	I	I
Choreoid, athetoid and dystonic.	0	I	I	2
Choreoid movements with tension.	0	0	I	I
Choreoid, athetoid and tension.	I	I	I	3
All types of involuntary movement.	I	7	9	I7.

voluntary movement in cases of dyskinesia, is due to actual paresis of movement, except in mildly affected patients. In the latter, paresis of movement is usually slight. But that some degree of paresis of movement is present is suggested by the fact that of the 17 cases in the series, 12 showed abnormality of muscle tone, tendon jerks or the plantar responses, in addition to their involuntary movements. The increase of tone was slight in 4 of the 7 cases in which it was observed, and it was severe in none of the cases. In 3 patients the increase of tone was predominantly rigid in 4 spastic or mixed. In 3 cases it was unilateral. In 2 it affected one arm and the opposite leg predominantly. The plantar responses were extensor on one side in 3 and bilaterally in 4 cases. These findings are probably to be regarded as manifestations of incidental cerebral damage additional to that found in the basal ganglia and their cortical connections. In all cases in the category of dyskinesia in the series the impairment of motor function was predominantly due to the involuntary movements which occurred on attempts at voluntary activity.

Some idea of the severity of the impairment of function is gained from an examination of what the children were able to do for themselves and to what degree the development of their motor functions were retarded. The average age at which unsupported walking was achieved was 2 years 5 months. Three cases aged 6 years, 9 years and 12 years were still unable to walk at the time of examination. The gaits of most of the other children were grossly abnormal. Five children over the age of 3 were unable to feed themselves completely. /

completely. Only 4 of the 17 cases were able to manage their buttons and laces, and thus dress themselves completely without help. Writing was extremely difficult for the majority of these children and was very clumsy and inadequate when it was possible. Three of the children over the age of 5 had speech that was so impaired that it was incomprehensible. Yet the intelligence of the children with dyskinesia was certainly not more severely impaired than in other forms of cerebral palsy. (Table 168)

Athetosis.

Athetosis occurred as the only form of involuntary movement in only one patient who showed moderately severe tetraplegic involvement of remarkable symmetry. The involuntary movements occurred only with voluntary activity in the limbs, and were more readily produced by attempts at fine movement of the fingers and hands than on movements at the proximal joints. They were produced in the arms by voluntary movements of the legs, however, and were slightly apparent in the opposite limb to that being used. Thus when an object was being grasped with the right hand, marked involuntary extension and separation occurred in the right fingers, associated with extension of the wrist and some pronation in the forearm. At the time, however, associated movements, of similar type, though less marked, were evident in the fingers of the opposite hand. When she walked the feet seemed to writhe a little and the gait was rendered clumsy. At the time some slow involuntary movements of the hands and wrists were apparent /

apparent bilaterally.

There was no apparent abnormality of muscle tone and the tendon jerks and the plantar responses were normal. Associated movements of extensor type were apparent in the limbs on strenuous voluntary activity. (Table 168)

Choreoid movements.

Choreoid movements occurred alone, without other involuntary movements in only one patient who showed slight tetraplegic involvement. She was a mentally defective girl, 13 years old at the time of examination. There was some generalised clumsiness of voluntary movements, probably not without normal limits. In addition she showed inconstant sudden involuntary movements of the limbs on voluntary activity of choreoid type. The movements were more marked when she was excited or under stress than when she was at ease. They varied in severity from day to day, and tended to be more evident towards the end of the day when she was tired than when she was fresh. The movements were more evident in the larger proximal joints than the smaller distal parts of the limbs. This resulted in the gait being somewhat jerky as the sudden movement put the leg transiently out of alignment and the whole child became stiff momentarily. In the upper limbs the movements were more liable to occur, voluntary activity necessitating more or less accurate portioning of the larger joints. Thus they occurred very frequently when the child was feeding, lifting food or a cup to her lips. The limb tended to be moved suddenly and uncontrollably with considerable violence, and any object in the hand might be

thrown /

thrown violently out of it. The ceiling of her home was marked by the food which had been projected on to it in this manner. (Table 170)

Athetosis and choreoid movements in association.

Athetosis and choreoid movements occurred more frequently in association than as isolated phenomena. In 6 patients they were the only involuntary movements which were apparent and in another 6 they were present in association with involuntary movement of different types.

The relative severity of the athetosis and choreoid movements in these cases varied greatly, as did the extent and severity of the movements encountered. Indeed, no case was very similar to any other in neurological findings.

Of the 6 patients showing athetoid and choreoid movements and athetosis, together with other involuntary movements, one was hemiplegic, 3 were triplegic, and 2 were tetraplegic. One was slightly affected, 3 moderately severely affected and 2 were severely affected.

(a). The choreoid and athetoid movements were modified, but not altered by the super imposition of other forms of involuntary activity, tremor, tension or dystonia. All 12 cases of choreoid and athetoid movements will be considered together therefore, and the ways in which other involuntary movements modified them will be discussed later.

(b). The movements were essentially similar to the involuntary movements described in the previous two sections, but the athetosis /

Case 197.

A boy aged ten years suffering from dyskinesia, in whom athetoid movements were the major cause of disability in some voluntary activities and choreoid movements in others.

The result of the third pregnancy to an obese mother aged 27 and her healthy husband. First child healthy. Second child died aged 9 months from bronchopneumonia. The patient was delivered precipitously at estimated 38 weeks' gestation after an uncomplicated pregnancy, with a birth weight of $5\frac{3}{4}$ lb. He breathed at once but was bright yellow in colour, and became progressively jaundiced during the next five days. Remained jaundiced for 6 weeks and was excessively drowsy from the second week until the third month. There was great difficulty in keeping him awake for feeds during this time and he vomited after almost every feed. At the age of three months he was admitted to hospital on account of feeding difficulties, but on discharge two weeks later he fed without difficulty. Suffered from recurrent bronchitis, diphtheria, measles, whooping cough, chickenpox and mumps before the age of three years.

He sat with support at the age of 9 months and walked with support at 22 months, unsteadily and falling a great deal. Walked without support rather unsteadily by the age of 2 years. Said his first words aged 18 months and had phrases by the age of two years. At this time drooling which had been constant before began to diminish, but "he seemed to be pulling faces whenever he talked". He always preferred his left hand but was observed to be clumsy in handling in play. He often knocked things off the table when he reached for them, especially with the right hand, and his fingers, especially on the left, did not appear to obey him. Though he was able to dress by the age of five he had great difficulty in getting his right arm into his jacket even at the time of examination and had only just learnt to manage buttons unaided. Though he had progressed moderately well at a school for physically handicapped children his writing was irregular and clumsy. He had to "write big" in order that his writing should be legible.

On examination he was a well built cheerful boy showing continual involuntary grimacing of the face and involuntary movements of the limbs. He appeared to have some hearing difficulty and was unable to hear a high-pitched whistle though he could converse intelligently. His speech was dysarthric, consonants being poorly articulated and the rhythm being slow and laboured. Intonation was monotonous. Speech was accompanied by much grimacing and at times by involuntary choreoid movements of the right arm which tended to assume a position of adduction and internal rotation at the shoulder, extension of the elbow, pronation of the forearm, and flexion of the wrist and fingers over the adducted thumb.

Apart /

Apart from inability to converge the eyes or look up, involuntary movements of the face on attempted smiling, and the presence of hearing impairment, cranial nerves were within normal limits. Power was fair on voluntary movements of all four limbs, but co-ordination of movement was impaired by the presence of involuntary movements and involuntary fluctuation of the state of muscle tone.

Involuntary movements were of three types. Brisk involuntary movements most marked in the proximal parts of the limbs which spread to involve the head and neck and to be associated with facial grimacing were present. They were present in all four limbs on attempted voluntary movement, but affected the right upper limb more than the left. Thus he tended to use the right upper limb extended at the elbow stiffly, and in spite of this when reaching for objects irregular involuntary movements would occur so that he was liable to knock things away before he could get his fingers round them. When he was excited the choreoid movement was almost flailing in type.

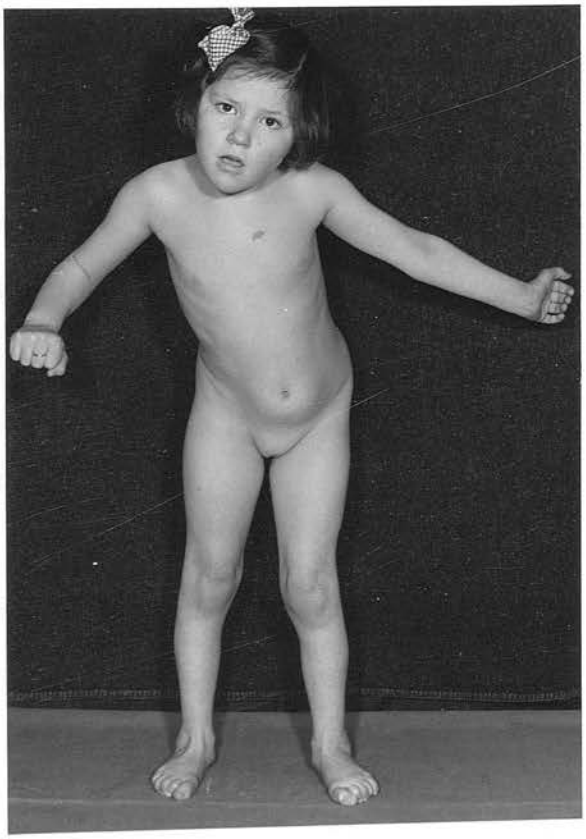
The second type of movement was more marked in the distal parts of the limbs and was slow and writhing in type. It was equally marked in both upper limbs, but more evident in the more frequently used and functionally better left hand. When he reached for objects or attempted to grasp them or make fine manipulations, his digits showed extension abduction and writhing irregular fluctuating activity spreading to the wrist and forearm over which he had no control for a few seconds, before his willed movement, usually of prehension or flexion of the fingers, was accomplished. This made fine activities very difficult for him, and though he could place the left hand in the position for doing shirt buttons the writhing athetoid movements of the digits, hand and wrist would frustrate his efforts for some time.

The third type of movement consisted of involuntary associated movements. Thus whilst he was using one upper limb, or exerting himself in any way the other limbs would involuntarily assume characteristic positions. In the upper limbs the typical position was of adduction and slight flexion at the shoulder, extension at the elbow, pronation of the forearm, flexion of the wrist and fingers over the adducted thumbs. The position was more readily assumed by the right upper limb than the left.

The tone of the limbs was increased even when he was apparently at rest, the increase being rigid in type and more marked in the right limbs than the left. The biceps, triceps, supinator, knee and ankle jerks were increased, more on the right than the left. The plantar responses were flexor when the head was turned away from the tested side and extensor when the head was turned in the direction of the tested foot. The abdominal responses were normal. No abnormal sensory findings were noted.

Figure 19 .

Precarious standing balance in a severely affected dyskinetic girl with choreoathetosis and dystonic features.



athetosis was modified by the choreoid movements to some extent and the latter by the athetosis. In general, the disturbance of function evident in the cases showing both types of movement was more severe than in those showing the movements not in combination. This was clearly seen in a number of patients in whom athetosis predominated in some limbs and choreoid movements in others. (Case 197).

The degree to which involuntary movements of choreoid spread to the trunk and to the other limbs varied greatly. in severe cases of involuntary movement, head, neck, trunk and all the limbs might be involved in involuntary movements elicited by voluntary activity in a single limb. Three patients showed marked involvement of the head and neck when voluntary movement of the more affected upper limbs was attempted.

Because of the severe impairment of all forms of fine voluntary movement which results from the combination of chorea and athetosis in the limbs, only 2 of the 17 patients were able to manage their buttons and shoelaces unaided. The writing of all was very poor.

Of the 12 cases, 10 showed associated movements of the contralateral arm similar to those found in hemiplegia and diplegic paresis. In 6 the movements were flexor in type and in 4 they were extensor in type. When the children were at rest little alteration of muscle tone was encountered, but the least excitement was liable to produce an increase of muscle tone and the appearance of involuntary choreoid movements. /

movements. Some increase of tone, usually slight, was evident at rest in 8 patients. In 3 the increase of tone was slightly rigid on one side and slightly spastic on the other. In 8 the tendon jerks were exaggerated, more on one side than the other in 3 cases, and more markedly in the legs than the arm in one case. Eight cases showed extensor plantar responses, 4 on only one side and 4 bilaterally.

Tremor.

Tremor of coarse type further impaired voluntary movement in 2 cases, one showing athetosis and in one case showing associated athetosis and choreoid movements. In the former case the tremor was evident only on voluntary movements, and appeared to involve the whole of the limbs undergoing voluntary movement, proximal as well as distal joints. In the latter case also the tremor was only evident on intention and was coarse in type. It affected the distal parts of the limb, the parts undergoing athetoid movement, rather than the proximal parts.

In both these cases there was quite a marked alternating increase and decrease in muscular tone in the affected limbs when the tremor was evident. The tremor was not present in limbs undergoing associated movements, the result of voluntary activity confined to other parts.

Dystonia.

Dystonic movement of the trunk and limbs was evident in 3 cases, one with choreoid movement and one with choreoid and /

Case 37.

Choreoathetosis and dystonia in a severely affected girl aged three years.

The first child born to healthy parents. The mother was aged 24 years at the time of delivery. Labour occurred at term after a normal pregnancy and lasted thirty hours. Delivery was spontaneous in a nursing home. The cord had been prolapsed for twenty minutes before the child was born.

The birth weight was 7 lb. 2 oz. and the child was severely cyanosed, limp and apnoeic. Resuscitation, involving injections into the cord and artificial respiration, resulted in breathing occurring after about two hours. Regular respiration was only established after 2 hours and 20 minutes. As a result of the resuscitation the child received burns of one leg from a hot water bottle.

The day after birth the child was whimpering and restless, with small range facial movements and intermittent twitching of the limbs. Her cry was high pitched. Twitching ceased after three days and thereafter, apart from a marked tendency to lethargy, the child appeared to be normal.

The child was eventually discharged from hospital at the age of three months, by which time it was evident to the mother that the child was not normal. The limbs were stiff, the shoulders tended to be adducted, the elbow to be extended, the wrists and fingers flexed. The legs tended to be held in an extended position and the feet to be plantar flexed. She moved the limbs very little and was very still when put down. She did not attempt to use the hands until the age of one year, and when she did so, the left was the first to be employed, the right not till three months later. When the hands were first used the movements were noted to be very jerky and clumsy. She could maintain the sitting position with support from the age of 15 months. She said her first words at the age of $2\frac{1}{2}$, and about the same age developed a great interest in picture books and passing traffic.

True involuntary movements of the limbs became apparent at the age of about six months. They resulted in the child suddenly becoming stiff, with the back arched and the neck hyperextended. The upper limbs showed shoulders adducted, elbows extended, the forearms pronated and the wrists and fingers flexed. The legs were strongly extended and the feet plantar flexed. This movement resulted from any sudden change in the position of the child or attempt at voluntary movement of the limbs. They persisted until the age of three years, but by this time the involvement of the trunk was less marked.

Twitching attacks of the limbs with impairment of consciousness which lasted for a few seconds became apparent at /

at the age of six months at the same time as attacks of dystonia became evident. These attacks of impairment of consciousness began with twitching of the eyes, of flickering type, and facial movements. The fingers worked bilaterally and the limbs became stiff, and the back arched. During the time the back was arched, the child's consciousness appeared to be severely impaired for perhaps half a second. The arching of the back and the stiffness of the limbs relaxed suddenly and was succeeded by a period in which the child felt flabby and consciousness was fully regained. Initially the attacks occurred two to three times a day, but at times when she was suffering from one of her periodic respiratory infections they were very much more frequent and occurred 25 - 30 times a day. The attacks became much less frequent at the age of 2½ years and were only occasional from the age of 3 years.

On examination she was a helpless, rather immobile child who could sit with support but rather unsteadily without it. She had about six words of spontaneous speech which were used appropriately but were very poorly articulated with much drooling, grimacing and associated involuntary movements of the face, neck and both upper limbs. She appeared to see normally and to hear. There was a bilateral convergent squint with a coarse nystagmus whenever she tried to fix her gaze. Her face was relatively immobile and any attempt to smile resulted in marked grimacing. She swallowed infrequently and drooled continuously.

Power in all four limbs was less than normal but not grossly impaired. She had no prehension in either hand, but a full range of voluntary movement was possible otherwise. All attempts at voluntary movement were grossly impaired by the presence of marked involuntary movements of three different types, and a tendency for her limbs to become very markedly rigid whenever she tried to use them. The first type of involuntary movement was choreoid in type and involved rapid changes in the position of the operating limb, most marked at the proximal joints. This resulted in flailing of the limbs and a tendency for them to be thrown into characteristic positions whenever she tried to move them. The position of the upper limbs was of adduction at the shoulder, extension of the elbow, pronation of the forearm, flexion of the wrist and fingers over the adducted thumb, and of the leg adduction at the hip, extension at the knee, plantar flexion of the foot and flexion of the toes.

The second type of movement, usually combined with the first, was a slower writhing movement of the fingers or toes, involving the hand or foot and wrist or ankle joint, of athetoid type. When she reached for objects, in addition to the jerking, flailing, irregular movement at the proximal joints, there would be these slower movements, usually with extension and abduction of the digits, extension of the wrist and pronation of the forearm in the distal parts of athetoid /

Figure 20 .

Severe dyskinesia.(Case 37)showing characteristically poor head control and associated involuntary movement of the left upper limb.....the "decerebrate"position of which is apparent.

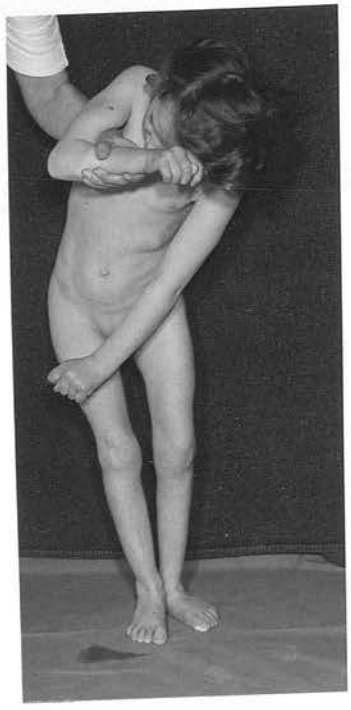
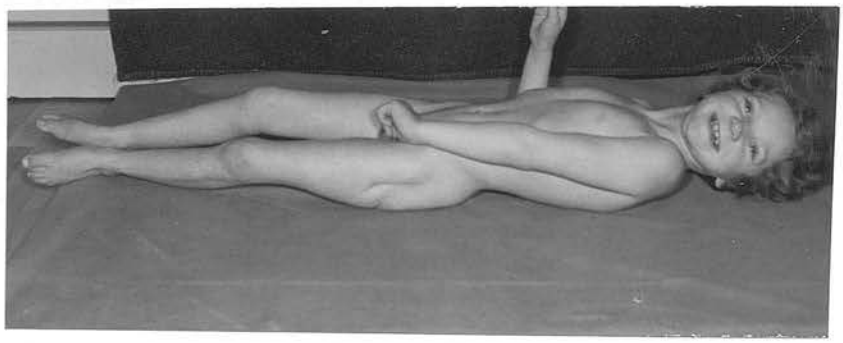


Figure 21 .

The tonic neck reflex in severe dyskinesia,(case 37). The upper limb towards which the head is turned assumes a "decerebrate" position,whilst the contralateral upper limb flexes and adducts at the shoulder,flexes at the elbow and pronates in the forearm.



athetoid type.

The third type of involuntary movement was a slow, very powerful movement of the proximal parts of the limbs. This occurred most often on changes of posture or in the limbs contralateral to those undergoing voluntary movement. They were dystonic in type and often of the nature of involuntary associated movements. When she tried to exert herself strongly or became excited these dystonic movements affected not only the limbs, but also the trunk, so that involuntary extension of the neck and back occurred, and what appeared to be fragments of a generalised movement towards the opisthotonic position could be seen.

At rest the tone of the limbs was within normal limits, but unless she was completely at rest any manipulation of the limbs resulted in a rigid increase of tone, rather more striking in the lower limbs than the upper. This increase of tone was also evident whenever she attempted any voluntary movement.

The result of the presence of involuntary movements and involuntary increase of tone as described was that practically all voluntary activities which she attempted were impossible. A particularly severe disability was that any change of posture, especially towards the erect position tended to result in dystonic movements of the limbs and upper trunk.

At rest the biceps, triceps, supinator knee and ankle jerks were within normal limits. The plantar responses were infantile in type. She appreciated pin prick in all four limbs but detailed sensory testing was impossible.

and athetoid movements. In all the cases the pattern of the movement was similar. Following a sudden choreoid movement of an upper limb, the limb would tend to take up a position of extension, the forearm pronated and the fingers flexed. The head and neck which had been involved to some degree in the chorea movements of the upper limb then tended to extend and the back and legs to follow suit. The opposite upper limb might or might not show extensor positions.

Synchronous with the involuntary dystonic positioning occurred a very marked increase of tone in all the limbs. The dystonic position was only apparent momentarily, but was extremely powerful and resulted in a complete disorganisation of voluntary activity. None of the patients exhibiting these severe movements could walk. All were classified as being severely handicapped. (Case 37)

It is interesting that two of the three patients with dystonic movements showed marked associated movements of the extensor type on voluntary activity of the limbs. Clearly there is some similarity between these cases and those in the dystonic stage of diplegia.

Tension.

Sudden generalised increase of muscular tension affecting equally agonists and antagonists was almost invariable in those showing choreoid movements when voluntary activity was attempted. The degree to which this tension affected limbs other than the one being moved was variable.

In /

In 4 cases, one with choreoid movements only, and 3 with athetoid movements also the tension was severe. In these patients voluntary movements were checked and sometimes rendered impotent by its occurrence, as in those with dystonia.

Other causes of marked increase in muscular tone in these cases were conditions of excitement, fatigue or mental stress. As with choreoid movements, tension was found to vary very greatly in its severity in the same patients at different times and in different conditions. Cases seen briefly at clinic examinations had, in fact, frequently been diagnosed as cases of generalised rigidity, whereas careful examination with the child at ease would have shown that, in fact, muscle tone at rest was normal.

It was interesting to observe that patients with marked increase of tone voluntary movement function was sometimes quite as much impaired by this as it was by actual choreoid movement of the limb. It seems much more likely that the increase of tone is a true reflex manifestation than that it is a conscious act on the part of the child to lessen his involuntary movements as has been suggested. (Phelps, 1943)

In one patient the tension on voluntary movement was evident in three limbs, and one arm was much less affected by it. In other 3 patients all the limbs showed the increase of tone though to different degrees. In 2 of the patients the state of tension was also found in the contralateral upper limb to that being used. In both extension of contra-lateral limb was evident. (Case 145).

<u>Other findings in the limbs</u>. /

Table 170.

Severity and extent of impairment of voluntary movement in 17 patients with dyskinesia.

	Hemiplegic.	Triplegic.	Tetraplegic.	Total.
Mild.	0	2	2	4
Moderately severe.	I	I	4	6
Severe.	0	2	5	7
Total.	I	5	II	17.

Other findings in the limbs.

Vasomotor changes similar to those found in hemiplegic cases were encountered in none of the patients classified as suffering from dyskinesia. The feet of some of the more severely incapacitated patients confined to bed or wheel chairs were found to be colder than the hands. Cyanosis was encountered in none of the cases.

Generalised dwarfing was not a feature of dyskinesia and the heights of children over the age of 5 appeared to be within the range of normal according to available figures. Asymmetry of the limbs was not marked. In only one case was one upper limb more than $\frac{1}{2}$ inch shorter than the opposite upper limb, and in 2 cases shortening of one-third of an inch was found, with less marked dwarfing in the corresponding lower limbs, in all three cases.

In no case was disturbance of superficial sensation or joint sensation noted. In view of the cases of hemichorea and hemiathetosis with hemi-anaesthesia recorded in the early literature this was something of a disappointment. (Charcot, 1868).

Speech defects in dyskinesia.

Speech defects occur frequently in patients suffering from dyskinesia for a number of reasons. The development of speech may be retarded because of mental retardation or impairment of hearing, and poverty of language and immature and distorted articulatory patterns will result. Incoordination of the lips, tongue and palate are the rule rather than /

Table 171.

Speech defects in 15 patients suffering from dyskinesia.

	No. of patients.
Articulatory defects only	3
Articulatory defects and deafness.	3
Articulatory defects and respiratory difficulties.	3
Articulatory diffic lties with respiratory difficulties and deafness.	I
Articulatory difficulties and stammer.	I
Rudimentary speech only.	4
Total.	15.

than the exception in dyskinesia and may result in complete inability to make intelligible speech even when the child is of average intelligence. Incoordination of the respiratory muscles also occurs frequently. It results in arrest of the flow of speech or in "explosive speech" in many cases. Since the causes of abnormal speech are commonly multiple in individual cases the analysis of speech disorders in dyskinesia may be extremely difficult. In Table 171 an attempt has been made to assess the most important causes of speech defects in the 17 dyskinetic patients in the series.

In 2 patients there was some incoordination of lip, tongue and palatal movements, but speech was intelligible before the age of five years and they are not, therefore, considered to have significantly defective speech, (Cases 54 and 68). The remaining 15 patients were not easily intelligible. Four, one of whom was very deaf and mentally defective (Case 194) had only rudimentary speech sounds and detailed analysis of speech was impossible. Hearing appeared to be intact in 3, one of whom was severely mentally defective and had a cleft palate (Case 202 and the others, extremely severe respiratory and articulatory incoordination (Case 37 and 178).

Dysarthria due to incoordination of the articulatory organs appeared to be the predominant cause of defective speech in 3 patients, and was present together with severely impaired hearing in another 3. In addition to articulatory and /

and respiratory incoordination one child was also very deaf, (Case 145). Articulatory and repiratory incoordination alone were present in 3 cases. One unfortunate child had articulatory difficulties and quite a marked stammer, the latter being familial, (Case 112).

Cranial nerve involvement in dyskinesia.

Visual impairment. Impairment of vision due to optic atrophy or field defects was not found in any of the patients with dyskinesia. One patient showed severe myopia and another severe astigmatic refractive error. Two cases showed inequality of the pupils, though these reacted normally to light and accommodation.

Eye movements. Strabismus was found in 6 cases. It was due to unilateral abducent weakness in two patients and to bilateral abducent weakness in four.

Nystagmus was evident in 5 cases. In all it was most marked when an attempt was made to fix the gaze, and two of the children, who were able to read, held the head and eyes in curious positions which they assumed in order to minimise the effect of the nystagmus. The eyes were held to the side to which the nystagmus was least severe and the head rotated to the opposite side. In 3 of those with nystagmus, strabimus at rest was apparent, in one unilateral abducent paresis and in 2 bilateral abducent paresis.

Facial involvement. Facial paresis of upper motor neurone type was found in 5 patients. Only in one was facial asymmetry /

present at rest. In the other 4 patients it was only manifest on movement. Facial overaction was encountered in one of these patients. In the other 3 there was facial lag on the affected side on emotional and voluntary movements.

Involuntary movements of the face, fluctuant or tremulous in type, occurred on voluntary facial movement in 6 patients. In 5 of these the involuntary movements were unilateral; in one they were bilateral. They were evident in all on speech, or attempts at speech as well as on movements elicited for the purposes of examination. In all these patients overflow of involuntary movement from the upper limb on the affected side to the face occurred. In some patients the involuntary facial movement was marked and associated with opening of the mouth and very bizarre facial expressions.

Hearing. High frequency deafness was confirmed by audiometry in 7 of the 11 cases of dyskinesia tested. The audiograms of 4 cases were normal. In 6 cases audiometry had not been performed.

High frequency deafness has been noted to occur frequently in cases of dyskinesia and is considered to be an important cause of speech defect. (Asher, 1952).

Swallowing. In 4 patients, all of whom showed dyskinesia of moderately severe or severe degree swallowing was impaired, and drooling was marked to the age of 5 or 6. In 2 of these patients swallowing occurred less frequently than once in two minutes on the average. All the children who showed difficulty in swallowing showed speech defects. Two had no comprehensible /

comprehensible speech, and the two others showed explosive speech and dyslalia. Three of the four patients with impairment of swallowing had facial involvement. Two had involvement of the tongue.

Impairment of voluntary movements of the tongue. Abnormality of the movements of the tongue was noted in seven cases. In all these cases voluntary movements of the tongue were impaired by involuntary movements, usually of a fluctuating character. Because of them the full range of voluntary movements was impossible and the child was unable to maintain the tongue even momentarily in any one position.

All those who showed tongue involvement also showed speech defects. In two speech was so poor as to be incomprehensible.

Intellectual impairment in dyskinesia. The schooling received by the 17 cases of dyskinesia and their intelligence as measured by different observers in different circumstances and by different tests are shown in Table 172 and 173 . It will be noted that of the five children with intelligence quotients of over 85 only two attended normal school. This indicates the severity of the educational handicap suffered by children with dyskinesia. It is interesting that the numbers of cases of normal intelligence who find their way to special school for those with cerebral palsy include a high proportion of children with dyskinesia. The severity of /

Table 172.

The intelligence quotients of 17 patients with dyskinesia.

I.Q.	115 plus.	110 plus	85 plus	70 plus.	55 plus.	Under 55.	Untested.
No. of patients.	1	2	2	4	2	1	5.

Table 173.

The schooling of 17 patients with dyskinesia.

School.	No. of patients.
Normal schools.	2
Schools for physically handicapped.	7
Schools for mentally handicapped.	2
Ineducable.	1
Not yet at school.	5
Total.	17.

of the educational disability suffered by these cases is out of proportion to their intellectual impairment.

The numbers of cases in this survey are insufficient for any opinion to be given as to the correctness of the impression that children with dyskinesia show less impairment of intelligence than do other types of cerebral palsy. (Dunsdon, 1952; Evans, 1948).

Overactivity. One intelligent child with triplegic dyskinesia of moderate severity showed overactive behaviour similar to that described when hemiplegic cases were discussed.

Epilepsy in patients with dyskinesia. Four patients showed epileptic phenomena. One patient showed petit mal beginning at the age of 6 months and continuing at the age of 2½ at the time of examination, 15 to 20 times a day. Three patients showed seizures of different types. (One of these, Case 37 has beem noted on page 731). In the latter the attacks are also rather similar to the so called striatal attacks found in some cases of encephalitis lethargica.

One patient, (Case 37) showed attacks lasting one minute to 80 seconds and occurring 15 - 20 times a day. In these consciousness was impaired but not lost. The attacks began with a sudden incoordinate movement of the head and neck, the eyes showed marked nystagmus and the left shoulder was moved rapidly and incoordinately from the trapezius.

No /

No abnormal movements of the legs were noted. After about one minute the attacks ceased and she seemed normal again. During the attacks the tone of the upper limbs was very greatly increased. The girl was a severe tetraplegic case. The attacks began at the age of 6 months and were still present at the age of 3.

One severely tetraplegic girl aged 6 (Case 202) started to have attacks at the age of one year and these were evident at the time of examination. Her attacks began with sudden severe impairment, but not absolute loss of consciousness, and a marked rigidity appeared in the limbs and trunk. The head and neck extended and deviated to the right. The upper limbs flexed briskly and sometimes hit her severely in the face. The lower limbs extended. After about one minute the child seemed normal. Sometimes she was incontinent during the attack.

Other findings in cases with dyskinesia. One patient showed bilateral congenital dislocation of the hips which it had not been possible to correct by orthopaedic means. One patient was stated to have shown "club feet and hands" immediately after birth, by the hospital notes. The feet had been energetically treated from the age of 2 days and at the age of 10 when she was examined, only slight talipes equino-varus deformity was evident bilaterally. Case 202 had a cleft palate as mentioned.

One patient, with kernicterus showed decaying green teeth. /

Figure 22.

Teeth in a choreoathetoid child who suffered from kern-icterus as a complication of haemolytic disease of the newborn. The line of enamel hypoplasia across the canines is well seen.

Figure 23.

Severe dyskinesia associated with mental retardation and cleft palate.(Case 202) At the age of six the child has no independent standing balance and shows marked dystonic movements in the erect position.

teeth. Five of the 7 patients still with their first dentition showed very decayed teeth. A history of very poor teeth in the first dentition was obtained in 6 of the other 10 cases. The finding of poor teeth did not seem to be related to the dyskinesia being due to rhesus incompatability or not.

ACQUIRED DYSKINESIA.

One patient developed involuntary movements of the limbs apparently following a head injury. A case summary is presented.

Case 125.

A girl aged 13 years with choreoathetosis developing after a head injury at the age of three years.

She was the result of the sixth pregnancy of a healthy mother, the father and four siblings being well. One sibling suffered from tuberculosis. No abnormality of pregnancy, labour, delivery or neonatal course was noted. Her motor and speech milestones were within normal limits.

At the age of 41 months she fell from a first storey window onto the pavement. She cried on hitting the ground and walked back unsteadily, staggering to the house without losing consciousness. Next day she was noted to be very drowsy and unresponsive. She had two black eyes and "was talking to herself in a daft kind of way". She was taken to hospital where X-ray of her skull showed no fracture, and she was discharged after two days' observation. She remained ratherdrowsy, had lost her appetite and complained continuously of headache. She seemed a little unsteady. Her black eyes disappeared after ten days, and she gradually became more normal in behaviour, though less enterprising and active than previously.

Three months after her accident involuntary movements of the right hand were noted when she tried to reach for objects or when she was playing. At times the movements merely prevented her from handling things neatly and were jerky in type, but at other times they were violent and would result in objects which she was holding being thrown into the air.

The /

The movements became more obvious and impaired her activities progressively during the next two to three months. They affected the left hand as well as the right. She became unable to dress herself or manage small manipulations which she had previously enjoyed. She could no longer feed herself without shaking food off her spoon or fork or putting it all over her face. Her walking became unsteady and as she walked "her arms seemed to be dancing". She seemed less alert than previously, said less and what she did say was less clearly spoken.

She was given a place in a school for mentally retarded children at the age of five years, her I.Q. being estimated at 54. She made very poor progress there, and writing and handwork continued to be impossible for her though she had learnt to dress herself and to feed herself rather clumsily by the age of nine years.

On examination she was a well built adolescent girl with a rather dull and expressionless face. She did not appear to have any defect of hearing or vision. She was very slow to comprehend what was said to her and obeyed commands very sluggishly. Her speech was slow and laboured, consonants being very poorly articulated, and it was difficult to make out what she said. Her sentences were simple in form. No abnormalities were noted in the cranial nerves.

Power of voluntary movement was quite good and there was a full range of movements at all joints. Co-ordination of all voluntary activities was poor, however, being impaired by choreoathetoid movements bilaterally, more evident on the right than the left and affecting both the proximal and distal parts of the limbs. It was difficult to distinguish an athetoid or choreoid component in the involuntary movements, for they appeared to spread from the proximal to the distal part extremely smoothly, causing a complicated elaborate mixture of shoulder, elbow, wrist and hand activity over which the patient had no control.

Associated contralateral movements of the upper limbs were extensor in type and were exaggerated. Tone at rest was slightly increased and rigid in type. The biceps, triceps, supinator knee and ankle jerks were slightly increased, rather more on the right than the left. The plantar responses were flexor. No abnormal sensory findings were noted.

When she walked she did so on a rather broad base with considerable tilting of the body, first to one side and then the other. The footsteps were irregular in timing and spacing, and there was some excessive irregular involuntary movement of associated type in both upper limbs.

It /

It is difficult to attribute the history and clinical findings in this case to any well recognised disease entity. The possibility that there may have been intracranial haemorrhage at the time of the head injury and that signs and symptoms of dyskinesia appeared as a result of secondary pressure or torsion effects must be considered. On the other hand choreoathetosis appears to be a very infrequent sequel of head injury.

Patients suffering from unclassified cerebral palsy.

Two patients suffered from cerebral palsy which not not be fully classified under any of the previously defined categories.

The first ,Case I02,was a moderately severely handicapped girl with severe arthrogryposis.She also had associated cranial nerve paralysis,Moebius' syndrome.There was bilateral paralysis of all external ocular movements including eye closure,though involuntary eye movements could be produced and the eyes made to close by "threat".There was bilateral paralysis also,of her face,but tongue and palate were normal.In addition to rigidity in the lower limbs there was also overlying spasticity with increased knee and ankle jerks and extensor plantar responses.She was mentallu handicapped;her intelligence quotient being estimated at 62 on the Terman Merrill scale,but was progressing reasonably satisfactorily in a school for physically handicapped children.

The second unclassified case was of a boy suffering from post-encephalitic Parkinsonism following an acute illness some three years before he was examined. During the period of the survey and for some time before and after it there appeared to be little if any change in his clinical picture which was one of generalised rigidty and tremor with little,if any intellectual impairment.Though his speech became so slow and slurred that it was almost unintelligible,and he could walk only very short distances without support his mind remained active.He was fully reported and attempts to treat him were described by Farquhar,I952.Following the completion of the survey this patient deteriorated rather rapidly within a period of about two years and died in I957.In view of this deterioration it is clear that he was not correctly classified as a case of cerebral palsy retrospectively,though the classification could be defended at the time it was made.